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THE PATIENT-PHYSICIAN RELATIONSHIP IN SICKLE-CELL DISEASE AT YALE-NEW HAVEN HOSPITAL

Douglas Theodore Fleming

Yale University

1991

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The Patient-Physician Relationship in Sickle-Cell Disease at Yale-New Haven Hospital

A Thesis Submitted to the
Yale University School of Medicine
in Partial Fulfillment of
the Requirements for the Degree of
Doctor of Medicine

By

Douglas Theodore Fleming
(1991)

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Dedication

This thesis is dedicated with great affection to:

- My wife, Robin Buckingham, the light of my life,

and to

- My parents, Betty and Robert Fleming, whose support for me through my life and education has been unfailing.

In addition, Robert Fleming's wonderful computer expertise and programming were invaluable in analyzing the data on hospital visits.

Acknowledgements

The most profound contribution to this project was made by the patients themselves. Their sharing of their experiences was most gratefully received.

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- Ms. Nancy Mason, whose excellent computer programming and good humor made possible the search of YNHH billing records

- Prof. David Williams, of the Sociology Department of Yale University, whose insights on the doctor-patient relationship pivotal in designing the study.

A note on abbreviations

The abbreviation "ED" is used throughout the text to stand for "Emergency Department". In some interviews, patients refer to the "ER", or Emergency Room, as the Emergency Department was previously known.

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I. Introduction

Sickle-cell disease presents a unique set of problems for the patient-physician relationship. From the patient's standpoint, sickle-cell disease represents a life-long, disabling chronic disease, which can lead to many psychosocial difficulties and dependence on analgesic medications. From the physician's standpoint, the disease represents a profound failure of modern medicine: despite exquisite understanding of the pathological basis of the disease in abnormal hemoglobin, no treatment has yet been found to substantially ameliorate the suffering caused by the disease, nor to halt progression of the disease in most cases. In addition, no objective method of diagnosing painful crisis yet exists, which places diagnosis largely in the subjective realm.

These inherent difficulties in the patient-physician relationship are exaggerated in the modern Emergency Department, in which a large volume of patients with increasingly acute illness burdens both physicians and patients. Furthermore, Emergency Departments are increasingly taxed as they become the means by which many patients seek primary care for relatively less acute illness. In this environment, patients who return repeatedly, requesting pain medications but not responding over the long term to any known treatment, run the risk of arousing suspicions of drug dependence or drug-seeking among Emergency Department staff and house officers. For the patient, the Emergency Department, especially in a teaching hospital, is an often chaotic place in which treatment is likely to be given by previously unfamiliar physicians.

One may hypothesize that in such an environment, a circle of suspicion and frustration may develop. This theoretical circle might begin with a small subset of sickle-cell patients whose disease has especially

severe manifestations, and who seek care frequently in the Emergency Department. Some of these patients may become well known to Emergency Department staff as requiring frequent and high doses of narcotic analgesics. Irrespective of the actual prevalence of drug dependence among patients, suspicions of drug dependence may arise, leading to a tendency among Emergency Department physicians to limit the prescription of narcotic analgesics to sickle-cell patients in general. Thus, many sickle-cell patients in crisis may receive less than the quantity of narcotic analgesics necessary to control their crisis pain. In such a situation, their requests for additional pain medication may lead to escalating suspicions of drug-seeking among ED staff and increased limitations on prescription of narcotics. In this way a circle of suspicion and frustration may begin to fuel itself.

The present study aims to investigate this hypothetical circle at several points, and to gather the information necessary to organize management interventions to interrupt it. To this end, the study will be arranged in three parts: 1) a literature review, 2) an analysis of hospital visits and admissions to YNHH during the period of a year, and 3) interviews with a subgroup of patients who were the most frequent visitors to the Emergency Department during the study year. In the discussion section, a number of possible interventions will be discussed in light of the results of the study.

II. Literature Review

The difficulties of treating patients with sickle-cell disease have been intensively studied ever since Herrick first definitively described the manifestations of the disease in 1910. In this section, therefore, an overview of the literature on sickle-cell disease will be given, concentrating on factors which influence the interaction between the patient with sickle-cell disease and his or her health care providers.

This section will begin with a general discussion of the various attempts by modern medicine to understand and treat sickle-cell disease. After all, the relationship between doctor and patient in sickle-cell disease has been conditioned to date by the intractability of the disease to all but symptomatic treatment. In addition, modern medicine's efforts to objectively confirm the presence of sickle-cell crisis will be reviewed. These efforts have been in three general areas: laboratory tests, physical examination, and psychological testing.

Next, the literature on the doctor-patient relationship in general will be briefly reviewed, with emphasis on the nature of the "sick role" assumed by many patients with chronic disease. In particular, "Social Labelling Theory", an area which concerns itself with the way in which medical judgements or diagnoses in themselves affect patients' lives and social interactions, will be discussed.

Literature on psychological aspects of sickle-cell disease, especially as regards the effects of chronic disease and pain on patients with sickle-cell disease, will be reviewed in some detail. Special emphasis will be placed

on "secondary gain" and adversarial relationships with health-care providers.

Finally, pain control during sickle-cell vaso-occlusive crisis will also be reviewed, concentrating on the role of narcotic analgesics and non-steroidal anti-inflammatory medications in sickle-cell disease.

A. Medical issues:

1. Frustration in treatment since discovery of the disease

a. Historical issues

A look at the history of sickle-cell disease in modern medicine reveals a good deal about the frustration felt by clinicians working with this disease every day. After all, though it is a history of triumphs in understanding the molecular basis of the disease, attempted treatments for the disease have been either toxic or ineffective. Therefore, this section will begin with a brief discussion of the discovery of the pathogenesis of the disease. ([1]).

Though syndromes now identifiable as sickle-cell disease had been recognized by African peoples for many years previously ([2]), the disease was first described to Western medicine by James B. Herrick, at a conference. His full report was published in the Annals of Internal Medicine in 1910 ([3]). Herrick's investigation had begun some six years

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[1] The review of this subject by Conley is the most extensive one available. see Conley, 1980

[2] see the reports of Felix Konotey-Ahulu, cited in Conley, p. 319

[3] Herrick, 1910

earlier, during an examination of the blood from an anemic student from the West Indies. After Herrick's initial report, the clinical pattern was established slowly, as case reports appeared over the next several decades.

In 1927 Hahn and Gillespie reported that sickling of the red cells occurs when hemoglobin is deoxygenated, a great leap forward in understanding the pathogenesis of the disease: "It is reasonable to suppose that sickle cells are formed in the body of an affected person wherever the oxygen tension and hydrogen ion concentration are such as to render the distorted form of the corpuscles stable" ([4]). In 1933, L.W. Diggs presented a remarkably complete description of the lesions seen at autopsy in a large group of patients with sickle-cell anemia, and suggested a mechanism for his observations: "A possible explanation of the capillary engorgement is that the elongated and spiked cells interlock and pass with more difficulty through narrow spaces than do normal cells." ([5])

A tremendous breakthrough in the understanding of the sickling process came during a conversation between William B. Castle and Linus Pauling, on a train ride from Denver to Chicago in 1945. Pauling recalls that Castle, in mentioning the birefringence of deoxygenated, sickled red blood cells seen with polarized light, suggested that sickle cells might "contain an abnormal kind of hemoglobin, which when deoxygenated has the power of combining with itself into long rigid rods, which then twist the red cell out of shape" ([6]). Pauling began to work on the problem with his graduate students, and in 1949 he described the altered electrophoretic

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[4] Hahn, 1927

[5] Diggs, 1934

[6] Pauling, 1955. Note that Castle is modest in his own account of the same conversation: "I am equally clear that I did not make the further generalization that it was orientation of the hemoglobin that might be doing this [ie. causing birefringence]" in Conley, p.339

mobility of the hemoglobin of patients with sickle-cell anemia, and ascribed the change to an alteration in the globin. With electrophoresis, the group was able to prove that the red blood cells of those with sickle cell "trait" contained both normal and sickle hemoglobin. Thus it was proven that the person with sickle-cell trait was a heterozygous carrier of the sickling gene.

It was in 1956 that Vernon Ingram combined electrophoresis and chromatography in examining tryptic fragments of sickle hemoglobin, to show that the abnormality lay in a single peptide. ([7]). In the years following this discovery there was a tremendous burst of research into the structure of hemoglobin, and by 1961 the entire primary structure of hemoglobin had been elucidated. Thus it was discovered that the site of the amino acid replacement in sickle hemoglobin is at the sixth residue of the beta chain. As Conley puts it, the "first molecular disease" had been discovered ([8]). After this, successes came quickly with the elucidation of the position of the aberrant residue on the surface of sickle hemoglobin, and with insights into deformation of the cell membrane by polymerized sickle hemoglobin.

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[7] Ingram, 1958

[8] Conley p. 346

b. Clinical manifestations of sickle-cell disease

The investigation of sickle-cell disease is made more difficult by the protean forms which sickle-cell disease may take in the individual patient. In addition, different clinical syndromes are experienced by patients with different combinations of the genes responsible for sickle-cell disease.

Patients who are heterozygous for the sickle gene (AS) possess one copy of the sickle gene and one copy of the gene for "normal" hemoglobin A. These individuals are said to have sickle-cell "trait", and generally experience minimal clinical problems. Their overall life expectancy and rate of hospitalization is the same as those of individuals homozygous for hemoglobin A. Because their red blood cells contain enough hemoglobin A to prevent sickling under most circumstances, they experience sickle-cell crisis only when extremely dehydrated or hypoxic. Many patients with the AS phenotype have impaired ability to concentrate urine, and a few have painless hematuria due to infarction of the renal medulla, which is especially vulnerable to sickling ([9]).

Patients who are homozygous for the sickle gene (SS) make no hemoglobin A, and may experience an extremely wide range of clinical manifestations, from mild to life-threatening, and in a variety of organ systems. Starting from about six month of age (when fetal hemoglobin has diminished enough to allow sickling by sickle hemoglobin), children with sickle-cell disease have a general delay in growth and development, and an increased tendency to develop infections, especially those due to encapsulated bacteria such as pneumococcus. The function of the spleen, which would normally remove such pathogens from the circulation, is

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[9] Bunn, 1987, p.1519

increasingly impaired by microinfarcts ([10]), until by adulthood only a nubbin of fibrous tissue remains.

Adults who are homozygous for sickle hemoglobin have a severe hemolytic anemia, with hematocrits in the 18 to 30 percent range. Mean red blood survival is 10 to 15 days, as compared to a normal survival of 120 days. Because these patients are so dependent on ongoing production of red blood cells, any infection which affects erythropoiesis, such as that of parvovirus, can result in aplastic anemia and a rapid drop in hematocrit. However, the most characteristic syndrome affecting patients with sickle-cell disease is the vaso-occlusive "painful crisis", which can attack various parts of the body, particularly the abdomen, chest and joints. About a third of painful crises are preceded by a viral or bacterial infection. Others appear to be precipitated by cold weather or by dehydration.

The extremely wide range of manifestations and complications of vaso-occlusive crisis makes differential diagnosis difficult. It is often difficult, for example, to differentiate between abdominal sickle crisis and some other acute process such as biliary colic, appendicitis, or a perforated viscus. Patients with abdominal crisis normally have no rebound tenderness and normal bowel sounds, and if surgery is performed, no gross evidence of infarction or ischemia is usually found. When sickle crisis appears in the lung, it may either mimic, or be complicated by, pneumonitis or pulmonary infarction. Similarly, when sickle crisis occurs in the extremities, it may be complicated by osteomyelitis, or confused with gout or rheumatoid arthritis. Patients with sickle-cell disease may even present with cerebral manifestations such as stroke, seizure or coma. In fact, vaso-occlusive crisis may affect almost any organ system, particularly the lungs, kidneys, liver, skeleton, and skin. ([11])

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[10] see Pearson, 1985, p.392

[11] See Bunn, 1987, p. 1520-21

Among the various other variants of sickle-cell disease, an especially important one occurs in patients who are doubly heterozygous for sickle hemoglobin and hemoglobin C ("SC disease"). These patients have a mild to moderate hemolytic anemia, and less frequent sickle crises. For unknown reasons, patients with SC disease are especially vulnerable to ocular complications, particularly proliferative retinopathy and retinal detachment, as well as hematuria from renal medullary infarcts, and avascular necrosis of the femoral head.

The protean manifestations of sickle-cell disease complicate the doctor-patient relationship in a variety of ways. Clearly, the possible presence of such complications as osteomyelitis or pulmonary infarction can make for an extremely complex and expensive diagnostic workup, which may nevertheless be completely negative. The choice of whether to begin such a workup is very difficult, and may have to be based on subjective criteria such as the patient's expressions of pain. In a patient suspected of simulating painful crisis in order to obtain pain medications, the diagnostic uncertainty is even greater. After all, even a patient who has shown irrefutable drug-seeking behavior in the past may still be at genuine risk for a fatal sequel of sickle-cell disease in the present instance.

c. Various attempts at treatment.

Treatment of sickle-cell disease has not been as spectacularly successful as research into its underlying causes. Herrick reported success with rest, nourishing food, administration of boric acid to leg ulcers, and

the use of iodide, arsenic, and thymol. ([12]). Other treatments in the first half of the century included splenectomy, liver diet, iron, blood transfusion and a wide variety of other measures, none of which proved to be more than transiently successful ([13]).

As the pathophysiology of the painful crisis began to be understood, various attempts were made at prophylaxis. The reader is referred to reviews by Charache and Conley ([14]) for a more detailed discussion of these attempts at prophylaxis. Some successes were achieved, as, for example, with partial exchange transfusions, which are still used to the present day. However, because of side effects such as long-term iron overload and the development of alloantibodies, the use of exchange transfusion is limited to preparation for surgery and other stressful situations, or the prevention of recurrent strokes. Similarly, oxygen was found to be somewhat effective, but carried the danger of depression of erythropoiesis and increased anemia ([15]).

Most of these attempts at treatment, however, were total failures, some of them spectacularly so. Nitrites and Carbon monoxide, for example, were used in the hope that sickling could be prevented by the resulting methemoglobin and carboxyhemoglobin, but these treatments could not be safely administered. Also tried were sodium bicarbonate and sodium citrate, in the hope that increased pH would decrease sickling, but could not be shown to be effective in controlled trials. Urea and Cyanate as potential anti-sickling agents both generated tremendous excitement in laboratories across the country, but proved to be either ineffective, toxic,

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[12] Herrick, 1910

[13] Charache, 1974

[14] Charache, 1974, pp. 698-705, and Conley, pp. 356-358

[15] Charache, 1981, p. 195-196

or both. Cyanate, in particular, caused a severe peripheral neuropathy, as well as cataracts.

Even at present, treatments in use for sickle-cell disease do not attempt to modify sickle hemoglobin at all, but rather, attempt to ameliorate symptoms by causing an increase in production of fetal hemoglobin ([16]). For example, some elevation of fetal hemoglobin has been achieved with the hematopoietic hormone erythropoietin, though the most recent study on its effects did not show any significant benefit ([17]). In addition, numerous cytotoxic drugs are known to increase production of fetal hemoglobin. For example, it has been shown that 5-azacytidine could stimulate dormant gamma globin genes in adults with sickle-cell disease, thereby increasing production of fetal hemoglobin. Similarly, in several studies with small patient samples, hydroxyurea has been shown to be effective in causing elevated levels of fetal hemoglobin over long periods with minimal toxicity, and with decrease in painful crises ([18]). However, none of these therapies has been yet shown to do more than reduce somewhat the frequency of painful crises; no known therapy can yet reduce the severity of crisis when it does occur. At present, medications such as hydroxyurea are considered experimental ([19]).

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[16] Platt, 1988

[17] See Goldberg, 1990. Goldberg et als' trial examined the effects of erythropoitin alone and with hydroxyurea in 5 patients.

[18] Charache, 1987. Recently Goldberg et al (1990) have achieved success in 5 patients in increasing fetal hemoglobin and decreasing the rate of hemolysis in sickle-cell disease. Their trial of hydroxyurea also included erythropoietin, which was not found to be beneficial.

[19] Ironically, one of the most important medications in use against sickle-cell disease, namely, penicillin, does not even affect sickle cell crises, but rather protects against infections of pneumococcal and other encapsulated bacteria in sickle-cell patients with decreased splenic function; that is, it "compensates for the splenic paralysis sickling causes". See Platt, 1988, p.1479

Thus, though promising treatments are on the horizon, treatment of sickle-cell anemia remains at present about where it has been for decades, with the mainstays of medical treatment being analgesia, oxygen, and intravenous fluids ([20]).

d. Programs to screen for Sickle-Cell Disease

Given the failure of any medical therapy so far to significantly alter the course of sickle-cell disease, it is not surprising that various attempts have been made to reduce transmission of the sickle gene. In fact, a number of mandatory screening programs for newborns and adults were begun in the early 1970's, in the hope that genetic counseling of carriers of the sickle gene would be effective in reducing the incidence of the disease ([21]). However, most of these programs were poorly organized, and for the most part offered little counselling or follow-up to people identified as carrying the sickle gene. Moreover, since prenatal diagnosis of the disease was not available at the time, "prevention" meant advising affected blacks not to have children, and was seen as having racist motivations ([22]). Within a few years, mandatory testing of black patients was on the wane.

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[20] Charache, 1974 and 1981, Platt 1988

[21] President's Commission, 1983, p. 21-22, and Stamatoyannopoulos, 1974, p.271

[22] Robinson, 1973, p. 324. Also note that the use of penicillin as long-term prophylaxis against encapsulated organisms in children did not begin on a wide scale until Gaston's report in 1986. See Platt, 1988.

2. Lack of ability to verify sickle crises

Another area of intense research has been the search for an objective test to confirm the presence of vaso-occlusive crisis. Considering the many neurological, hematological and psychological factors which may influence the pain of vaso-occlusive crisis, as well as the many medical manifestations of crisis, it is not surprising that no test has been found which is both sufficiently sensitive and sufficiently specific for routine clinical use ([23]). There are three general areas in which research has attempted to objectively differentiate the pain of "genuine" vaso-occlusive crisis from "non-organic" pain, namely, laboratory examination, physical examination, and psychological examination.

a. Laboratory tests for Vaso-occlusive Crisis

Put briefly, there is no laboratory test to verify the occurrence of sickle-cell vaso-occlusive crisis. However, several recent attempts have been made. For example, Brownell has described the use of serum ferritin levels as an indicator of sickle-cell crisis ([24]). The most obvious difficulty with this approach is that, as an acute-phase reactant, ferritin levels rise in a wide variety of circumstances. In an attempt to overcome the limitations in specificity of laboratory tests taken singly, several writers have employed combinations of measurements to confirm sickle-cell crisis. For example, increases in percentages of dense cells, erythrocyte sedimentation rate, fibrinogen levels, Magnetic Resonance Imaging and serum prostanoids have all been used, in various combinations, to verify sickle-

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[23] Shapiro, 1989, p. 1031. Note also that it is still unclear what determines severity of the disease in a given patient -- on this topic see Schechter, 1982

[24] Brownell, 1986

cell crisis ([25]). None of these has been successful, and none is routinely used in clinical practice.

b. Physical Examination for Vaso-occlusive Crisis

Clinical methods to objectively distinguish "genuine" vaso-occlusive crisis from "non-organic" pain have not been described specifically for use in sickle-cell disease. The discernment of the organicity or non-organicity of pain by physical examination would typically be considered in the realm of "clinical intuition" ([26]). For example, "It is widely thought that pain that never fluctuates in intensity of pain in a nonanatomic distribution suggests the involvement of psychologic or learning processes" ([27]). However, hard evidence for such views is lacking.

Work to clarify this issue has been done in the case of low back pain, by Waddell. In his 1980 study, Waddell ([28]) did not directly address the issue of "organicity", but rather tested the ability of various findings in the physical examination to predict the response of a patient with low back pain to physical treatments or surgery. For example, the examiner may try

1) rolling the skin of the patient over the lumbar spine, or

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[25] Lawrence, 1987 and Mankad, 1987

[26] Bonica, 1990, p. 1457

[27] Bonica, 1990, p. 597-98

[28] Waddell, 1980

2) lightly placing one's hand on the patient's head or rotating the patient's upper body (shoulders and hips together).

Since none of these maneuvers increase pressure in the lumbar spine, complaints of pain by the patient on these maneuvers raise the suspicion that the pain cannot be ascribed to a structural lesion in the spine. Other findings include

3) a significant discrepancy in pain between straight leg raising in the seated and supine positions,

4) non-physiological regional disturbances of sensation, distribution of pain, or weakness, and

5) over-reaction to the test stimulus, with excessive verbalization, grimacing, or other "excessive" pain behaviors.

Waddell showed that those patients with three or more positive findings rarely responded to any physical treatment or to surgery. It is important to remember, as Bonica points out, that "these are not tests of malingering; they only indicate that the patient's pain behaviors cannot be ascribed solely to structural lesions of the spine" ([29]).

The situation in sickle-cell disease is different than that of low back pain in several respects. First, the pain of vaso-occlusive crisis may occur in almost any area of the body, making specific "positive" findings on physical examination difficult to describe. Second, in sickle-cell disease there is no modality (as, for example, surgery in some kinds of low back

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[29] Bonica, 1990, p. 1458

pain) successful enough in treating the underlying disease that patients' response to it could be correlated with given findings on physical exam.

c. "Objective" Psychological tests

Many attempts have been made to employ supposedly "objective" psychological testing to identify those patients who are prone to "non-organic" pain. For example, elevations in several subscales of the Minnesota Multiphasic Personality Inventory, especially Hypochondriasis, Depression, and Hysteria, have been noted in chronic pain patients ([30]). These scores have been interpreted to mean that chronic pain patients are characteristically preoccupied with their illness, unhappy, lacking in psychologic insight, and tending to develop somatic symptoms in response to stress and conflict ([31]). However, no single, pathognomonic pattern is observed on MMPI among patients with chronic pain, nor, of course, can it be established which, if any, of the chronic pin patients has an organic cause for the pain.

A number of "objective" psychological indicators have been used to assess psychological factors in sickle-cell disease, but none has been successful in predicting clinical features of the disease in a given patient, nor in characterizing "genuine" vaso-occlusive crisis. Psychological testing of patients with sickle-cell disease will be discussed under "Psychology" below.

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[30] Fordyce, 1976

[31] Bonica, 1990, p. 602

So far, we have reviewed the three main areas in which research has attempted to objectively verify the presence of vaso-occlusive crisis -- in the laboratory, the examination room, and the psychologist's office. The failure of these efforts has a profound effect on the doctor-patient relationship, since it means that physicians cannot objectively differentiate "true" sickle-cell crisis pain from pain of other causes, including factitious pain in the context of drug-seeking. As with other conditions in which the principle complaint is chronic pain, there is no "gold standard" for proving "genuine" sickle-cell vaso-occlusive crisis.

The literature on "objective" verification of pain often phrases the problem in terms of the patient, that is, it attempts to identify patients whose pain is non-organic. Waddell's system for identifying patients who will not benefit from surgery is one example. However, such an approach applied to patients with sickle-cell disease may give misleading results, because the same patient who may fabricate the symptoms of a sickle-cell crisis one week may have a "genuine" crisis the following week. In other words, both "untrue" and "true" crises may occur at different times in the same patient. Therefore, the "validity" of sickle-cell pain must be judged anew for each sickle-cell crisis.

B. The Doctor-patient relationship in Sickle-cell Disease:

1. General considerations

The patient with sickle-cell disease seeking care in the Emergency Department of Yale-New Haven Hospital faces a large system developed to evaluate and treat efficiently a great number of patients. Consequently, the relationship between individual doctor and patient must be viewed in terms of its position within that larger system. Therefore, it will be useful to discuss the work of David Mechanic, who has examined the effects of the "bureaucratization" of medical care in large medical institutions.

Mechanic points out the inherent conflict in the Emergency Department situation between the need to be efficient and the desire to provide care for patients' social and emotional needs ([32]). Bureaucracy, after all, "encourages specialized activity, routinized procedures and modes of operation, [...] and there is a strong tendency for bureaucracies to try to limit client control." ([33]) In addition, as medical institutions become larger and more impersonal, a variety of intermediaries become interposed between the patient and the professionals who control their care. In fact, patients may even have difficulty ascertaining who is actually responsible for their care ([34]). Thus patients may feel that they are facing an impersonal system that does not understand their individual problems or pain.

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[32] Mechanic, p. 414

[33] Mechanic, p. 416

[34] Mechanic, p. 416

Conversely, Mechanic points out that within a bureaucratically organized medical system, physicians may feel equally frustrated. A special source of difficulty for the physician is the "patient with a high inclination to use medical care" ([35]) but no objectively verifiable basis for his or her symptoms:

> "There is no end to the depth of exploration that is theoretically possible, and the doctor knows that he must live with the fact that it can never really prove that there is no underlying physical basis. [...] At some point he must begin to consider the risks of continued exploration: the dangers of the various diagnostic procedures, the economic costs of continued searching, and the probability that continued searching may reinforce the attitude of the patient that he is indeed physically sick."
>
> ([36])

Mechanic points out that physicians may in many cases feel that the treatment they provide may have at best a problematic value for their patients. Certain patients may develop an overdependence on physicians, and may actually be "adversely affected by particular kinds of intensive care and attention". ([37]). For example, "excessive focus on symptoms and life difficulties may reinforce an already hypochondriacal pattern, induce or encourage further displays of illness behavior, and bring about reduced coping effectiveness." ([38]). In addition, such patients may be thought to be exploiting the "sick role", with its exemption from social responsibilities

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[35] Mechanic p.426

[36] Mechanic p.426

[37] Mechanic, p. 431

[38] Mechanic p. 431

and obligations ([39]). Physicians may thus suspect that this exemption from responsibilities is one more aspect of the "secondary gain" patients may derive from their illness.

2. The "Social labelling" Perspective on Illness

In studying the treatment of patients with sickle-cell disease, it is essential to remember that most patients with the disease come in with the diagnosis of sickle-cell disease, and that physicians will bear this in mind in all subsequent medical decisions. In addition, the "illness label" of sickle-cell disease in itself can have profound effects on patients' lives, irrespective of the symptoms of the disease at any given time. In this context, it will be useful to discuss the sociological literature on "social labelling", which describes the way in which medical judgements or diagnoses in themselves affect patients' lives and social interactions.

An overview of "Social Labelling Theory" can be found in a 1981 review by Nancy E. **Waxler**. She suggests that

> "A person is labelled as 'ill' in the course of social negotiations between himself, his doctor, his family, and sometimes ward staff and others. The outcome of such social negotiations is influenced by each person's beliefs and training and also by the social and organizational contexts in which the negotiation occurs. Once labelled as 'ill', the individual may find himself caught in the midst of a self-fulfilling prophesy". ([40])

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[39] see esp. Parsons, 1951

[40] Waxler p. 85

Thus, for example, a diagnosis of mental retardation 'labels' a patient in the eyes of many as having a profound disability, and may profoundly change that person's social relationships and future possibilities. However, as suggested in a 1973 study by **Mercer**, some parents, especially middle-class white parents, manage to "negotiate" another diagnosis to keep their children in normal classes, or simply send their children to a private or parochial school. Thus, the diagnosis of mental retardation ended up being disproportionately applied to black and hispanic students. ([41])

Similarly, individual patients may choose to adopt or discard a label of "illness", based not only on symptoms, but also on the patient's social and economic background. A good example is found in a 1968 study, by **Ludwig and Adams**, of patient cooperation in a general rehabilitation center. It was found that among all physically disabled people in the program, the ones most likely to drop out before completion of the program, against physicians' advice, were caucasians, men, the middle aged, and those with previous employment. Conversely, those more likely to complete the program were blacks, women, the very old and very young, and the unemployed. Thus, the data appear to suggest that those who were in the most powerful social position, and those who had employment to return to, were most able to declare themselves "normal" by dropping out of the treatment system. On the other hand, those in a less powerful social position, and those with no employment, seem to have had more reason to continue (or less reason to discontinue?) their "sick role" in treatment. ([42])

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[41] Mercer, 1973

[42] Ludwig and Adams, 1968

Characteristics of the institution also influence the application of the label of "illness". For example, the desire of a given institution to admit a patient formally (and thereby substantiate the patient's "illness") may be determined by the number of beds available at the time. In a 1975 study, **Harris** found that increases in the number of hospital beds in 56 counties in New York State led to increases in the number of patients in the hospital, not because of any increased need, but because physicians responded to the new, empty beds by increasing admissions and lengthening hospital stays ([43]). Similarly, the financial situation of a hospital, and the expected reimbursement from the patient or his insurance company, may influence the decision whether the patient is "sick enough to be hospitalized" ([44]). A patient with no insurance, for example, might conceivably be less likely to be admitted than a patient with insurance for the same illness.

The arguments of the "social labelling theory" can be applied in different senses to sickle-cell disease. Certainly, the presence or absence of sickle hemoglobin (hemoglobin S) is not "negotiated", and refutes the more extreme "social labelling theorists", who seem to imply that the labelling process "creates" the disease ([45]). In fact, the irrefutable presence of hemoglobin S in the red blood cells of patients with sickle-cell disease unavoidably conditions the entire interaction between an affected patient and his or her physician.

However, the presence or absence of vaso-occlusive crisis is "negotiated" in a very real sense, since it is usually entirely subjective, experienced by the patient but not verifiable by the physician. It is in this sense that "social labelling theory" can be most easily applied.

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[43] Harris, 1975

[44] see Waxler, p. 93

[45] see Waxler, p.99,note 2

3. The "Gomer phenomenon"

In this context, it is important to note that institutions may spawn a different, less formal sort of "label" for patients, not known to the patient, nor, for that matter, well known to the world outside the health care system. The term "GOMER" is a classic example, which is discussed in a 1985 article by **Leiderman and Grisso**. This term may have a special relevance to the care of patients with sickle-cell disease, whose care presents many difficulties.

First popularized in the novel House of God by Shem, "Gomers" appear as patients who frustrate the housestaff's every effort to ameliorate their condition. Leiderman and Grisso studied the way the term is used in an actual hospital by comparing a group of 18 "gomers" (as identified by housestaff) with a matched control group of "non-gomer" medical patients.

In Leiderman and Grisso's study, the image of the "gomer" which emerges is not a simple stereotype: "Gomer is neither a class-linked nor race-linked pejorative term, nor is the term applied merely to malingering, self-destructive patients" ([46]). However, "gomers" distinguished themselves from medical patients in the control group by the special frustrations which their illnesses and management posed for resident physicians: "Gomers suffered irreversible mental deterioration; their illnesses were complex and intractable; they were unable to resume normal adult social roles; and they had no place to go after discharge" ([47])

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[46] Leiderman and Grisso, p. 294

[47] Leiderman and Grisso, 1985, p.294. See also p.290, table 2

Such patients have special importance to the resident physician: "The lingering presence on acute medical wards of patients for whom modern medicine can provide no therapy, or frequently even diagnosis, challenges physicians' ideals about the powers of medicine" ([48]). In addition, "Gomers" rarely offer the resident physician an "interesting case", that is, one with educational value to the physician in training ([49]). Leiderman and Grisso suggest, however, that the situation may be different for the attending physician, who is "protected from the frustration, helplessness, and uncertainty that these patients generate". In fact, senior physicians have been known to severely censure housestaff for the use of epithets like "gomer" ([50]).

Leiderman and Grisso's article has the potential for application to patients with sickle-cell disease. With their multiple medical problems, sickle-cell patients require considerable effort by the housestaff, who are heavily involved in their care. Their illness is complex and intractable, and often is so disabling that normal adult social roles are impossible. Nevertheless, sickle-cell disease permits a relatively long life, and patients return again and again with frequently similar, inscrutable pain. As described above, few therapeutic options are available beyond analgesia and hydration. Response to therapy is most often slow and incomplete. Neither are patients with sickle-cell disease often "interesting cases". In short, patients with sickle-cell disease fulfill many of the criteria for the role of "Gomer".

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[48] Leiderman and Grisso, p. 294

[49] Leiderman and Grisso, p. 295

[50] Leiderman and Grisso, p. 295

4. The "Undesirable Patient"

In considering the nature of possible conflict between the patient with sickle-cell disease and his or her physician, it will be useful to have a framework for categorizing issues which lead to conflict. A useful article in this context was written in 1970 by Solomon **Papper**. He discusses the various types of "undesirability" which can make a patient unpleasant to his or her physician. ([51])

In Papper's scheme, qualities of patients "undesirable" to physicians can be broken down into five general groups:

1) Socially undesirable: this category includes, for example, alcoholic, dirty, uneducated, very poor patients, as well as patients belonging to certain ethnic or religious groups.

2) Attitudinally undesirable: for example, patients who "think they know too much", or who otherwise questions the physician's authority, or generally, those whose attitude frustrates the physician: "The problem here is the patient's lack of knowledge of, or failure to meet, the physician's expectations or needs" ([52]).

3) Undesirability on "physical" grounds: includes patients who lack a diagnosable physical illness or who do not respond to treatment (for example, "crocks"), and patients who have chronic (especially malignant) or simply "ordinary" (ie not "interesting") diseases.

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[51] Papper, 1970, p.259

[52] Papper, p.260

4) Circumstantial undesirability: for example, the patient who arrives late, or on a busy clinic day.

5) "Distraction" undesirability: for example, the patient who does not fulfill criteria for inclusion in the physician's research study, or who distracts him from his research.

Papper's scheme will be helpful in considering the nuances of conflict between doctor and patient. In the case of sickle-cell disease, patients may fit into more than one category of "undesirability", as seen by their physicians.

C. Psychological issues

1. Introduction

Psychological issues in the care of patients with sickle-cell disease are pivotal. After all, patients with sickle-cell disease are susceptible to all the difficulties of having a chronic, often painful, unpredictable disease with many ramifications for patients' occupational and social lives. Therefore, in this section a brief overview will be given of the literature on psychological issues in sickle-cell disease. Then will follow a more detailed look at six leading articles on the subject, written from 1976 to 1989.

The pain of sickle-cell vaso-occlusive crisis can be overwhelmingly stressful to patients. In fact, one writer, Barbara Shapiro, has compared the

intense, unpredictable pain of sickle-cell crisis to that experienced in the "learned helplessness" phenomenon in animals ([53]).

For all the pain they experience, children do not seem to become seriously "psychosocially impaired". In fact, children with sickle-cell disease tested with various psycho-social instruments seem, with a few differences, to be remarkably similar to healthy children of similar age and socio-economic status ([54]). Evidence seems to indicate that difficulties in psychosocial functioning seem to increase during adolescence, especially in young males ([55]).

Similarly, adolescence appears to be the time when adversarial relationships are most likely to develop between the patient with sickle-cell disease and his or her physicians and nurses ([56]). Puberty, difficult enough for healthy young people, may be especially difficult for both the patient and his health care providers. Barbara Shapiro, for example, has pointed out that adolescents with sickle cell disease must work through the usual developmental issues, which are marked by "intensive and often perplexing limit-seeking" ([57]).

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[53] Shapiro, 1989, p. 1032

[54] Kumar, 1976, discussed below and Lemanek, 1986

[55] Leavell, 1983, discussed below, and Hurtig, 1986

[56] Vichinsky, 1982, discussed below

[57] Shapiro, 1989, p. 1033

Going to the hospital for treatment of sickle pain is a learned behavior, which is influenced by a number of variables, including the patient's age, experiences during previous hospitalizations, home environment and support, and economic situation ([58]). Thus, it is not surprising that there is wide variation patients' propensity to use hospital services. Some patients are quick to visit a doctor at the onset of pain, while others attempt to manage even the most severe crisis at home. So, it is important to remember that the frequency of use of emergency services and the number of hospitalizations may not reflect the actual severity of a patient's illness.

Adults with sickle-cell disease appear to have a greatly increased prevalence of psychiatric and psychosocial impairment when compared to patients with otherwise similar socioeconomic status and severity of chronic illness, as discussed below ([59]). Patients with sickle-cell disease have been compared to patients with diabetes ([60]) and with asthma ([61]) (both discussed below).

A more detailed discussion of six leading articles on psychological aspects of sickle-cell disease will now follow.

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[58] Shapiro, 1989, p. 1033

[59] Damlouji, 1982 and Leavell, 1983, both discussed below

[60] Damlouji, 1982, discussed below

[61] Bobo, 1989, discussed below

2. Psychological issues: a more detailed look at six articles

a. **Kumar**, 1976

Santosh **Kumar** et al ([62]) studied a group of 29 adolescent children with sickle-cell disease, ages 12-18 years, selected apparently randomly from among patients making visits to the hematology clinic at the Los Angeles County/University of Southern California Medical Center (the article does not make clear how this sample was chosen). There was also a control group of 26 children drawn from a local high school, of the same age range and ethnic background. The subjects were given a battery of tests, including the "General Anxiety Scale for Children", the "Piers-Harris Self-Concept Scale", and the "California Test of Personality".

Results of these tests indicated that patients with sickle-cell disease are

> "no different from the comparison group without sickle cell anemia on personal, social, and total adjustment scales. [...] The patients show significantly higher tendencies to withdraw (be lonely) than the comparison subjects on the personal adjustments. The patients also had significantly lower social skills (liking for people, inconveniencing oneself to be of assistance to others, being diplomatic in one's dealing with both friends and strangers) than did normal subjects. The mean score for self-concept was significantly lower than that of the comparison group." ([63])

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[62] Kumar, 1976

[63] Kumar, p. 862

Interestingly, patients with sickle-cell disease scored lower on the anxiety scales than did normal students.

Kumar et al conclude that though patients with sickle-cell disease who have had consistent family support and health care are not different from a matched peer group in adjustment, as judged by the tests given. However, patients showed a lower "self-concept", and thus might benefit from "guidance and especially reassurance to strengthen self-confidence" ([64]).

b. **Barrett** et al, 1988

Drue H. **Barrett** et al ([65]) studied a group of 89 adult patients with sickle-cell disease being seen at a sickle-cell center at a hospital in Georgia. Barrett et al used the "Chronic Illness Problem Inventory" (CIPI) to assess psychosocial impairment in a range of areas, and also collected data on the number of acute emergency visits per month.

Results of the "CIPI" showed that patients rated as most severe their problems with employment, followed by sleep, eating, and financial difficulties. In addition, patients reported difficulty in the areas of Body Deterioration, Activities of Daily Living, and Assertion. The lowest problem severity ratings were found in the scales of Physical Appearance, Cognition, and Marital Difficulty. Men scored higher than women on 17 of the 18 scales, though not to a statistical significance of $P < .05$. Interestingly, there did not appear to be a correlation between scores on the "CIPI" and utilization of emergency services, years of sickle-cell disease, or age of the patients.

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[64] Kumar, p. 862

[65] Barrett, 1988

Barrett et al conclude that the studied population of patients with sickle-cell disease was having significant psychosocial distress, especially in the areas of employment, sleeping, eating, and finances. as well as in other areas. Although depression was not specifically measured, the authors speculate that their data may imply that "depression may be prevalent among this population" ([66]). They suggest that vocational rehabilitation and job training programs might benefit these patients.

c. **Leavell** et al, 1983

Sandra R. **Leavell** and Charles V. Ford ([67]) studied 16 adults with sickle-cell disease, including 8 men and 8 women, between the ages of 19 and 53 (mean age 32), recruited in an unspecified fashion from two university hematology clinics in Nashville. The authors used a structured interview to assess mood, personality, education and background in the patients, and also employed the "State-Trait Anxiety Inventory" (STAI) and the "Schedule for Recent Events" (SRE). The "SRE" yields "Life Change Units" (LCU's) as a measure of recent life changes and stresses. The authors also attempted to contact the patients six months later to ascertain whether they had experienced symptoms of sickle-cell disease in the interim.

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[66] Barrett, p.749

[67] Leavell, 1983

Results from the interviews and tests indicated that there was no consistent "specific characteristic personality or behavior problem associated with sickle-cell disease". However, 8 of the 16 patients were diagnosed with a "psychiatric disorder", including among other diagnoses sociopathy, history of substance abuse with hallucinations, drug-seeking, and depression. In particular, "psychiatric impairment" was greater in men than in women. Medical complications experienced by patients before the study were also more severe with males than with females. In addition, those patients who had had high stress (increased LCU's) in the previous 6 months, and those with low social support, were more likely to report having experienced symptoms of painful crisis in the 6-month follow-up period ([68]). Finally, "State-Trait Anxiety Inventory" scores for the 16 patients were not significantly different than those reported earlier for general medical patients ([69]).

Leavell and Ford conclude that their data did not suggest a "specific characteristic personality or behavior problem associated with sickle-cell disease (such as manipulative, narcotic-seeking behavior), but rather a wide range of different types of personal adjustment to the disease" ([70]). Regarding the lower levels of both medical complications and psychiatric symptoms among women, the authors speculate that "women with sickle-cell disease who survive to adulthood are less disabled and therefore have a more `normal' adjustment to life in spite of their disease" ([71]). Finally, they note that life stress, as well as lack of social supports, appears associated with increased symptoms of sickle-cell disease.

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[68] See Leavell, 1983, p.28-29

[69] Leavell, p. 25-29

[70] Leavell, p.29

[71] Leavell, p. 29.

Note that the authors do not give figures on how many of their female patients had had children. One wonders what effect pregnancy, which in sickle-cell disease has a high rate of complications, might have had on the authors' assessment that psychiatric morbidity was less in their female patients.

d. **Damlouji**, 1982

Namir F. **Damlouji** et al ([72]), compared 30 patients with sickle-cell disease with an age-, sex- and social-class-matched group of 30 patients with diabetes mellitus, all of them black, aged 15-56, from inner-city neighborhoods, recruited from patients seen at the Sickle-cell and Diabetes Clinics at The Johns Hopkins Hospital. The authors administered the "Social Functioning Schedule" (SFS) in a semi-structured interview, and the "General Health Questionnaire" (GHQ) to all patients in the study. In addition, the authors counted the number of medical complications noted in the charts of both groups of patients.

Damlouji et al found that both sickle-cell disease and diabetes mellitus were associated with social disability and psychiatric morbidity. Overall, the patients with sickle-cell disease had more difficulties than the patients with diabetes mellitus in 4 of the 12 scales of the "Social Functioning Schedule" (employment, financial problems, child care, and spare-time activities). There were no significant differences noted between the scores of men and of women. Overall, in both groups of patients, the authors note that although social maladjustment (ie "SFS" scores) and emotional disturbance (ie. "GHQ" scores) correlated with one another,

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[72] Damlouji, 1982

they did not directly correlate with the number of medical complications ([73]). Neither could age, marital status, nor educational level be shown to be associated with social maladjustment or emotional disturbance.

Damlouji et al speculate that factors which might have contributed to the social disability and emotional disturbance tested in their patients with sickle-cell disease and diabetes mellitus might have included "chronicity of illness, lifelong and chronic dependence on the medical care system, and the fact that both sickle cell anemia and diabetes are incurable". The authors suggest that the greater difficulties reported by patients with sickle-cell disease might have its basis in the fact that "the frequency and severity of painful crises are both unpredictable and out of the patients' control" in sickle-cell disease ([74]).

They also conclude that social maladjustment and emotional disturbance may be independent of physical condition (as judged by number of complications). However, their argument on this point is weakened by their failure to define how they counted "medical complications". Did they, for example, count "functional asplenia" in the same way as they did "cerebrovascular accident"? No clue on this point is given in the article.

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[73] Damlouji, 1982, p. 931

[74] Damlouji, p. 931

e. **Bobo** et al, 1989

Loretta **Bobo** et al ([75]) studied a group of 108 patients from the inner city of New Orleans, ages 15 and up, 48 of which had sickle-cell disease, and 60 of whom had asthma, as well as 50 of both groups' unaffected siblings. Bobo's group interviewed the patients by telephone, and employed selected scales from the Rand Health Insurance study to assess the patients' attitudes about prior and current health, health outlook and worry/concern, resistance or susceptibility to illness, as well as rejection of the sick role and attitude about doctors. In analyzing the data collected, the authors looked both at each of these groups separately, and at the aggregate of patients with sickle-cell disease and with asthma (ie the "chronic disease group").

Bobo et al found that health perception scores were significantly lower for the chronic disease group than for their unaffected siblings on all scales. In addition, the patients with sickle-cell disease reported significantly poorer health than the asthma patients in half the scales, including "health outlook", "resistance/susceptibility", and "sickness orientation". Both the patients with sickle-cell disease and patients with asthma reported high ratings of their own attempts to "reject the sick role" (eg keep going, fight their disease), comparable with their siblings' ratings.

When asked to give their opinions of the medical care they received (in terms of access, continuity, financing, humaneness, quality of care and satisfaction), both chronic illness groups, as well as their healthy siblings, gave largely similar ratings overall. The only statistically significant differences among the groups were significantly lower ratings given by sickle cell patients in comparison to ratings by asthma patients in two of the scales: humaneness of care (59 vs 65) and satisfaction with care (58

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[75] Bobo, et al, 1989

vs. 64). However, patients with sickle-cell disease did not report any more difficulty with access to health care, or with the costs of medical care, than did the patients with asthma or the group of health siblings.

Bobo et al give several reasons why patients with sickle-cell disease might report lower levels of health perceptions and opinions of medical care in several scales. Given that patients did not report more difficulty with access to health care than the asthma patients or healthy siblings groups, Bobo et al speculate that "These unfavorable ratings may have been influenced more by the chronic disease than the impact of services" ([76]). Thus, factors such as the recurring, unpredictable pain crises and early mortality of sickle-cell disease may have contributed to differences in the experiences of patients with sickle-cell disease and with asthma.

However, the authors mention another factor possibly contributing to negative opinions of medical care in this case, namely, the recent discontinuation (for lack of funding) of a special clinic for patients with sickle-cell disease ([77]). Patients who had received comprehensive care at the clinic were suddenly forced to rely on the emergency department and outpatient clinics of the public hospital. Unfortunately, the study by Bobo et al. did not address specifically in interviews with patients what effect the discontinuation of the clinic had on their views of their own health and of the health care system ([78]).

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[76] Bobo, p.11

[77] Bobo, p.12

[78] Bobo et al cite an article which is helpful in considering this question. Nicole Lurie and colleagues studied the effects on patients of termination from California's Medi-Cal program for indigent adults. The same study instrument, from the Rand Corporation Insurance Study, was used by Lurie's group, to demonstrate a fall in health perceptions after termination of Medi-Cal benefits. See Lurie, 1984 and Lurie, 1986

f. **Vichinsky**, 1982

Elliot Vichinsky and colleagues have done particularly useful research into the psychology of the doctor-patient relationship in sickle-cell disease ([79]). In examining the conflicts surrounding the treatment of sickle-cell disease at their hospitals in Oakland and Berkeley, California, Vichinsky et al found that housestaff felt "marked anxiety surrounding the evaluation of pain" in patients with sickle-cell disease. Conversely, the authors found dissatisfaction among patients with the care they received during sickle-cell crisis.

Reviewing the records of painful events requiring narcotic analgesia (a total of 1,295 painful events) in 152 patients in the previous year, the authors located the 31 patients of this group with the most crises and evaluated each one of them with a psychiatric examination. This preliminary data suggested that "depression, anxiety, and neuroses were frequently present in the chronically ill sickle cell disease patient" ([80]). Among other findings, psychiatric examination showed that "25 patients (80%) had clear-cut characterologic neuroses, including depression, hysteria, and hypochondriasis" ([81]).

In addition, Vichinsky et al studied the records of 101 patients receiving narcotic analgesics at their institutions (they do not clarify how this sample was chosen) in an effort to identify narcotic-related complications. The authors found that 6 patients had had "major motor seizures precipitated by large doses of demerol", 3 patients attempted suicide by drug ingestion (one died), 7 patients had "drug dependence", and

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[79] Vichinsky, et al, 1982

[80] Vichinsky, 1982, p.329-30

[81] Vichinsky, p.330, table 2

3 had "drug addiction" ([82]). No clarification is given about how these categories were defined in the chart review.

Interestingly, of the 1295 painful events reviewed, all initially thought to be sickle-cell crisis, reviewed by Vichinsky et al, 31 episodes (ie 2%) were subsequently found to have had other causes. Of these, 6 proved to be cholecystitis, 5 to be metastatic pancreatic cancer ([83]), 4 to be deep vein thrombosis. 2 episodes proved to be due to acute narcotic withdrawal ([84]).

Vichinsky et al conclude that depression, anxiety, and neuroses are prevalent among patients with sickle-cell disease. After their initial review of the psychology of patients with sickle-cell disease in their institutions, the authors instituted a comprehensive program to coordinate pain management in sickle-cell patients. This program is discussed below under "Treatment of Sickle Cell Disease in the Emergency Department".

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[82] Vichinsky, p. 330, table 3

[83] It is not made clear in the article whether these 5 episodes of pain from metastatic pancreatic cancer were in the same patient

[84] Vichinsky, p. 330, table 4

3. Psychological issues: conclusions

The six studies reviewed above are quite disparate in their approaches. Though five of the six studies employed at least one scalar (ie quantifiable) measure of personality, adjustment or psychological well-being, not one of these scales was employed in more than one study. In addition, patient samples and definitions of terms were different in different studies. Therefore, comparison of results among the six studies is difficult.

Age appeared to have a strong effect on psychological morbidity in sickle-cell disease. Whereas Kumar's sample of adolescents with sickle-cell disease was similar to a matched control group in terms of general adjustment and other measures, psychiatric morbidity in adult patients in other studies was prevalent. These adult patients reported difficulties with sleep, financial problems and body deterioration (Barrett), social disability and emotional disturbance (Damlouji), low health outlook, sickness orientation and dissatisfaction with medical care (Bobo). Patients in two studies reported that problems with employment were especially severe (Barrett and Damlouji).

In two studies discussed above, sickle-cell patients were found to have greater psychological impairment than their peers with other chronic diseases, including diabetes mellitus (Damlouji) and asthma (Bobo). The authors cite a number of possible reasons for this marked psychological impairment in sickle-cell disease relative to other chronic diseases, including the early onset of illness in sickle-cell disease, the frequency and severity of painful crises, and the early mortality of sickle-cell patients. Perhaps most important, patients with sickle-cell disease have very little control over their illness, as painful crises thus far are both unpredictable and unpreventable. This is in contrast to diabetes mellitus, which can be largely controlled (at least in the short term) by insulin and diet, and to

asthma, in which bronchodilators and other therapies can markedly reduce morbidity.

Two of the studies (Leavell and Vichinsky) employed structured interview and psychiatric exam to assess psychiatric health of sickle-cell patients. In Leavell's study, 8 of 16 patients were found to have a psychiatric disorder, including sociopathy, history of substance abuse with hallucinations, drug-seeking and depression. In Vichinsky's study, 25 of the 31 patients with the most crises were found to have characterological neuroses, including depression, hysteria and hypochondriasis. Thus, though both Leavell and Vichinsky found psychiatric morbidity prevalent in their patients, neither found any specific type of psychiatric difficulty which was singular to patients with sickle-cell disease.

In investigating the relationship between the patient with sickle-cell disease and his or her physician, one may ask whether or not the psychological stresses and impairment of sickle-cell disease predispose patients to dependence on pain medications. Unfortunately, the literature on the psychology of sickle-cell disease is not clear on this point. Though psychiatric morbidity appears common, it is so diverse in nature that no common thread leading to drug dependence can be identified. The topic of drug dependence and its prevalence among sickle-cell patients will be discussed further below.

D. Analgesics

The literature on the treatment of sickle-cell pain is relatively broad, and considerable disagreement exists about the value and dangers of different classes of analgesics in treating the disease. Therefore, a brief overview of the literature on pain relief in sickle cell disease will be presented here, concentrating on the use of narcotic analgesics, especially in the Emergency Department setting.

In dosing analgesics for patients with vaso-occlusive crisis, an understanding of the pharmacokinetics of various analgesics is pivotal. As Vichinsky points out, "duration of action of analgesia" refers to the period during which the patient experiences analgesia. This period is not necessarily the interval of adequate pain control ([85]).

Peripherally acting oral analgesics have a role in mild or moderate pain of sickle-cell disease. For example, acetaminophen is a commonly used agent, especially in children ([86]). In addition, aspirin and other non-steroidal anti-inflammatory drugs may be used in children who are afebrile and without signs of infection, and may be especially helpful in bony pain ([87]). In one controlled study, the non-steroidal anti-inflammatory agent diflunisal reduced pain towards the end of vaso-occlusive episodes, but not during the period of most intense pain at the beginning of the episode ([88]).

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[85] See Vichinsky, 1982, p. 331

[86] Pearson, 1984

[87] Shapiro, 1989, p.1034

[88] Perlin, et al,1986

For more intense pain, however, more powerful, narcotic analgesics are often necessary, starting with oral agents. On the weaker end of the spectrum of narcotic analgesics is oral codeine, which is often prescribed with a peripherally acting analgesic ([89]). Codeine would most commonly be prescribed for outpatient management of moderate pain ([90]).

Stronger opioids which can be given orally include hydromorphone, oxycodone, methadone, and immediate and sustained-release morphine. These oral medications may be used even for the management of severe vaso-occlusive crises ([91]), with an analgesic effect comparable to that of parenteral medications. Use of oral narcotics has been described in the Emergency Department, but admission may still be necessary for proper adjustment of dosage of monitoring of the patient ([92]). Nausea and vomiting may limit the usefulness of the oral narcotic agents. In addition, poor absorption of morphine may limit its use as an oral agent.

If admission and treatment with parenteral medication is necessary in vaso-occlusive crisis, morphine is considered an especially good choice ([93]). However, if untoward effects such as pruritus, nausea and vomiting occur and are persistent, another agent such as hydromorphone may be chosen.

Though meperidine is often prescribed for management of severe vaso-occlusive crisis, a number of dangers are associated with its use in

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[89] Shapiro, 1989, p. 1034

[90] Pearson, 1984, and Vichinsky, 1982

[91] Friedman EW, 1986

[92] Friedman EW, 1986 and Shapiro, 1989

[93] Vichinsky, 1982, p. 331 (table 5) and Shapiro, 1989, p.1035

patients with sickle-cell disease ([94]). Meperidine has a long-acting metabolite (normeperidine) which has only slight analgesic activity, and has been associated with excitation of the central nervous system, dysphoria, and seizures ([95]). Excretion of normeperidine into urine is decreased when urine is alkaline, which could lead to accumulation of this metabolite in the renal tubular acidosis of sickle cell disease ([96]). In addition, in one study meperidine levels in patients with sickle-cell disease were lower than those in control patients, though both groups had received the same amount intramuscularly ([97]). Thus, meperidine, at least when given by the intramuscular route, may be less effective in relieving pain in sickle cell patients than in other patients.

For patients in vaso-occlusive crisis who need parenteral analgesia, Vichinsky recommends a fixed dosing interval for morphine of 2.5 hours (at .15 mg/kg), to provide a constant level of analgesia. After the first 48 hours, the dose can usually be tapered. Once the dose of parenteral morphine has been lowered to half the initial dose, Vichinsky recommends switching to an equianalgesic dose of oral narcotics given every 4 hours ([98]).

Cole and colleagues report on the use of continuous intravenous narcotic infusions for the treatment of painful crisis in children ([99]). They studied a total of 98 painful crises in 38 patients. Of the 98 episodes of painful crisis, 76 were treated with continuous infusion of opioids

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[94] Shapiro, 1989, p. 1035

[95] Tang, 1980

[96] Szeto, 1977, and Shapiro, 1989

[97] Abbuhi, 1986

[98] Vichinsky, 1982, p. 331

[99] Cole, 1986

(morphine or meperidine) given on a standard protocol, and 22 were treated with intermittent intravenous bolus injections of meperidine. The authors found that patients given continuous infusions required more narcotic to control their pain and had more side effects than those treated with bolus injections alone ([100]).

Side effects of continuous intravenous narcotic infusions reported by the authors included nausea and vomiting, lethargy, and abdominal distention, as well as a significant incidence of chest syndrome, and severe respiratory distress in three patients. They note that "narcotic withdrawal or addiction was not observed" ([101]). Despite the many ill effects seen with continuous opioid infusion, the authors conclude that this modality is safe and provides adequate pain relief during vaso-occlusive crisis.

Patient-Controlled Analgesia (PCA) has been tried on a very limited basis for patients with sickle-cell disease. For example, Schechter et al have reported on a preliminary trial of PCA with adolescent patients, in preparation for a randomized controlled clinical trial. ([102]). Their sample consisted of only 3 patients, aged 12, 11, and 17 years old. The authors found that doses of morphine required for pain control were higher than expected. However, morphine requirements decreased on successive days as the pain crisis resolved, which was taken by the authors to imply that the drug was not being abused ([103]).

The authors argued that use of PCA broke the cycle of undermedication of sickle-cell pain, in which continued pain leads to a

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[100] Cole, 1986, p. 1255

[101] Cole, 1986, p. 1255

[102] Schechter et al, 1988

[103] Schechter et al, 1988, p.109

"seemingly undue interest in medication" by the patient, leading to suspicion of drug-seeking by the physician and continued undermedication. Interestingly, the authors believe that "deeply entrenched attitudes" in the medical profession toward patients with sickle-cell disease led to the refusal of one hospital to participate in a clinical trial of PCA for patients with sickle-cell disease ([104])

It is important to note that even if Patient-Controlled Analgesia is not available, many of the same advantages can be gained with a flexible dosing schedule ([105]).

Because there are no prospective, controlled studies comparing opioids and routes and schedules of administration in sickle-cell disease ([106]), it is difficult to make a definitive judgement on the efficacy of one versus the other. The reader is referred to the literature cited above for more detail on this subject.

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[104] Schechter et al, 1988, p.112. It might be noted that at the particular hospital cited by Schechter et al, PCA was already an established modality for treatment of post-operative pain.

[105] See Angell, 1982, p.98-99 and Shapiro, 1982, p.1036

[106] Shapiro, 1989, p. 1036

E. Narcotics addiction in patients with sickle-cell disease

Because "addiction" to narcotics is defined in so many ways, it is very difficult to compare figures on addiction among different authorities in the literature. However, because this issue is so important to the study at hand, an attempt will be made here to survey the literature on this subject. Generally, authors range from those offering personal impressions of the frequency of addiction to those who give statistics, but none has clearly defined the criteria for classification of a patient as "addicted". Thus, it could be argued that assessments of the addictive potential of opioids cited here reflect primarily the range of physicians' attitudes on the subject, rather than "hard data".

There is a wide spectrum of opinion on the addictive potential of opioids. At one end of the spectrum are those who feel that addiction in patients with sickle-cell disease is rare indeed. One writer from Great Britain, for example, Brozovic (1986), maintained that "not one of the 610 Brent, Camberwell, Haringey, and City and Hackney patients with sickle cell disease is known to be a drug addict" ([107]). In fact, this author felt that

> "to suggest that the pain of trunk crisis can be alleviated by reassurance and paracetamol and that administration of opioid analgesia leads to drug addiction and dependence pushes us back to where we were ten years ago when every patient with sickle cell disease was regarded as either a malingerer or a drug addict."
>
> ([108])

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[107] Brozovic, 1986, p. 624

[108] Brozovic, 1986, p. 624

Cole et al (1986) suggest that the withholding of analgesics may actually encourage addictive behaviors in children:

> "Inadequate pain management may ultimately lead the patient to seek pain relief by other than accepted means, a practice that could ultimately lead to addiction. Morphine or meperidine given in the hospital for a period of several days should not - and in our experience does not - result in drug dependence."
>
> ([109])

Cole et al note that no "addictive behaviors" were observed in any of their patients, nor were symptoms of withdrawal.

Other writers express similar views, but with less conviction, as in this quote from Heller (1988):

> "We are reasonably sure that these patients take the [narcotic] drug only for this purpose [of pain control] and are not addicted, and we are confident that the drug does not end up on the street market."
>
> ([110])

This author describes a group of frequent users of the Emergency Department, around whom the question of addiction appears to arise:

> "There are a few patients with such severe pain that they require Demerol compound, Dilaudid tablets, or other narcotic drugs. They are known to medical staff, come to the emergency department at frequent intervals, and are often

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[109] Cole, 1986, p. 1258

[110] Heller, 1988, p. 703

> hospitalized, and the risk of addiction can be disregarded in relationship to the obviously excruciating pain and severity of their disease".
>
> ([111])

The author concludes that "the need for pain relief is stronger than our fear of addiction."

Other writers express considerably more concern about the addictive potential of narcotics. For example, Charache (1981) evinces a high degree of suspicion about drug-seeking behavior:

> "Since drug-seeking is learned behavior, the physician must control the dose and time of administration of narcotics rather than give the patient the opportunity to seek the drug and have that behavior rewarded. There are some addicted sicklers; in my experience they all became addicted outside the hospital, and most admit they are addicted."
>
> ([112])

Charache does not make clear his criteria for addiction, nor does he clarify the meaning of his remark that patients become addicted "outside the hospital".

Vichinsky et al are singular in quantitating the risk of opioid addiction in their patients. In reviewing the records of 101 patients, apparently both adults and adolescents, for major and minor complications of narcotic use in sickle-cell disease, the authors uncovered 6 patients who had seizures from large-dose demerol, 3 suicide attempts (by overdose with

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[111] Heller, 1988, p. 703

[112] Charache, 1981, p. 196

narcotics), 7 cases of "drug dependence" and 3 cases of "drug addiction" ([113]). Of those classified as "drug dependent", none experienced withdrawal symptoms after counseling and elimination of narcotics. It is less clear how "drug addiction" was defined ([114]). For this reason, it is uncertain how the authors' estimate of the prevalence of "drug addiction" (3%) is to be compared with other studies.

Thus it appears that there is little data on the prevalence of addiction among patients with sickle-cell disease treated with narcotic analgesics. More information is available on the issue of addiction in patients with other illness characterized by pain, for example, chronic pain syndrome, or systemic illness such as malignancy. As this very broad field, the reader is referred to other articles on these topics ([115]).

F. Treatment of sickle-cell disease in the Emergency Department

Treatment of sickle-cell disease in the Emergency Department presents many challenges. These challenges have been discussed in a number of articles, three of which will be reviewed in some detail here. In one article, the type of pain relief given to children and to adults with sickle-cell disease visiting Emergency Departments is compared. In addition, two articles describe the authors' experience with the use of management protocols for treatment of sickle-cell disease in the Emergency Department.

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[113] Vichinsky, 1982, p. 330, table 3

[114] Vichinsky, 1982, p. 330, p.330

[115] See for example Bonica, 1990, p. 598, Brodner, 1978, Ready, 1982, and Portenoy, 1986

1. **Selbst and Clark**, 1990

Selbst and Clark sought to determine whether children with acute, painful conditions were treated in a similar fashion as adults with similar conditions. To this end, they examined the treatment of pain in sickle-cell crisis, lower-extremity fractures, and second- or third-degree burns, in 119 children treated at Children's Hospital of Philadelphia ED, as well as in 162 adults treated for the same conditions at the Medical College of Pennsylvania ED, over a seven-month period. Charts from these institutions were assessed retrospectively for patient diagnosis, disposition, and use of narcotic and nonnarcotic analgesics while in the ED as well as after discharge from the ED.

Fifty-three of the patients whose charts were reviewed by the authors had sickle-cell disease, including 27 children (defined as patients under 19) and 26 adults (defined as patients from 19-65). No patient over 65 years old had been treated for sickle-cell crisis. Patients with sickle-cell crises complicated by fever, stroke or other problems were excluded from the analysis. Analgesics prescribed were almost exclusively narcotic.

Selbst and Clark found that whereas all of the adults with sickle-cell crisis (26/26) had received pain medications in the ED, only 74% of the children with sickle-cell crisis (20/27) had received medication for pain, leaving 7 children who had received no pain medication at all. This difference in prescription of pain medication in the ED to children and to adults was significant to $P < .05$. However, differences in the prescription of analgesics on discharge from the ED to children was not significantly different from that to adults. Overall, for all three acute painful conditions examined by the authors, children received analgesics significantly less frequently than adults did.

The authors speculate that reasons for the less frequent prescription of analgesics to children are varied, and include fear of drug addiction, concern about respiratory depression, misconceptions about analgesia needed by children, and the more specific and vehement requests for pain medication by adults. The authors note that the prevalence of addiction among their adult patients is unknown, and that physicians' suspicions about addiction may have influenced the amount of narcotics they prescribed ([116]).

While useful, Selbst and Clark's study is somewhat limited by its retrospective nature, and its failure to analyze differences in prescription patterns between the two institutions, one of which (CHOP) serves primarily children.

2. **Powers**, 1986

Robert Powers has reported on a study of the use of individualized management protocols for treatment of sickle-cell pain. In the study, conducted in the Emergency Department of the University of Virginia Medical, 5 black men between the ages of 25 and 33 years were identified as "chronic ED users". These patients were advised that standard ED management of pain would consist of oral hydration, oxygen, and oral meperidine or methadone. Parenteral narcotics were given only at the request of the primary physician, or if the patient required admission for pain control ([117]).

Individualized treatment protocols, developed in concert by a primary physician and a hematologist responsible for the patient, were kept on hand in the Emergency Department, along with logs of ED visits. It is

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[116] Selbst, 1990, p. 1012

[117] Powers, 1986, p. 267

important to note that the "protocol for pain was begun only after history, physical examination, and appropriate laboratory testing excluded true sickle cell crisis, infection, or serious co-existent illness" ([118])

In the year following institution of the program for the five patients, Powers documented a decrease in the number of ED visits (from 163 to 56), as well as mean length of stay in the ED, percentage of visits given IM meperidine, and mean dose of IM meperidine. After informal surveys both of staff and of patients, Powers concluded that the program had been a success in reducing ED usage, in improving care, and in minimizing "confrontations between sickle-cell disease patients and professional staff".

The usefulness of Power's article is somewhat limited by the small size of his sample (5 patients). In addition, it should be pointed out that misgivings must arise from Power's apparent belief that "true sickle cell crisis" could be excluded by "appropriate laboratory testing" ([119]). In fact, as noted above, exclusion of "true sickle cell crisis" was a precondition for initiation of the treatment protocol. The implication seems to be that the protocol was used only on patients with proven drug addiction or drug-seeking behavior. Thus it appears that a fundamental misunderstanding of the limitations of diagnostic testing in sickle cell disease was at the heart of Power's study.

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[118] Powers, 1986, p. 267

[119] Powers, 1986, p.267

3. **Vichinsky**, 1982

Another study which examined the effects of a management protocol for sickle cell disease on utilization of Emergency Department services was conducted by Vichinsky et al. After doing a survey (described above) of the prevalence of depression and other psychiatric complaints in sickle-cell patients, the authors instituted an intensive multidisciplinary treatment program for patients with sickle-cell disease. The program included counselling and group therapy, vocational rehabilitation and school tutorial programs, and a standard protocol for medicating patients in both the inpatient and outpatient settings.

Vichinsky et al identified 10 patients (age range 19 to 41) who "fit into the chronic pain syndrome category", and examined their utilization of the Emergency Services and hospitalizations in the 6 months before and in the 6 months after institution of the program. The authors found a 58% decrease in Emergency Department visits and a 44% decrease in pain-related hospitalizations. They conclude that their multidisciplinary approach can "significantly reduce morbidity in sickle cell disease" ([120]).

The chief limitation of Vichinsky's data on ED visits is his vague description of the inclusion criteria for patients. It is unclear how the "chronic pain syndrome category" is defined. Thus, it is difficult to judge how such a program would affect ED use by sickle-cell patients in general. In addition, it is unclear whether the decrease in utilization of emergency services reflects a decrease in morbidity from the standpoint of the patient. Nor is it clear whether patients are seeking their care at another Emergency Department.

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[120] Vichinsky, 1982, p.332

4. Emergency Department Treatment -- an overview

Clearly, research into the treatment of sickle-cell crisis in the Emergency Department has been relatively sparse. What emerges most clearly from a number of articles on the subject is a sense of anger and helplessness, as in this quote from Power's article:

> "Frustration and anger often develop in ED staff as they are repeatedly confronted with patients demanding narcotics for relief of pain, which may be unaccompanied by objective signs of illness."

([121])

The patient with sickle-cell disease who has frequent painful crises may be considered together with other frequent users of Emergency Department. These patients, labelled "frequent flyers" in one article ([122]), are a source of special frustration for Emergency Department staff. However, little is known about this small but costly group of patients. Several studies have shown that frequent visitors to the Emergency Department tend to have a high incidence of chronic psychiatric conditions, alcoholism, and recurrent medical problems ([123]).

Pierce (1990), in a retrospective study of patients in Memphis who returned to the ED within two days of initial registration, found that patient-related factors (that is, in which the reason for return was "primarily the fault of the patient") accounted for 53% of return visits. Note that in Pierce's study, repeat visits by patients with sickle-cell disease

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[121] Powers, 1986, p. 267

[122] Schuckman, 1989

[123] Pierce, 1990, and Purdie, 1981

were lumped under "illness-related return visits", as an example, like asthma, of a "recurrence of a chronic disease process" ([124]), rather than under "patient-related visits", as drug-seeking and narcotics addiction might have been.

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[124] Pierce, 1990, p. 755

G. Comments on the literature review and its value for the present study

The literature review above has touched on a number of aspects of sickle-cell disease which present unique problems for the physician patient relationship. Not only can painful crisis not be diagnosed on any objective basis, presently available treatments are merely symptomatic. In fact, physicians may feel that treatment of sickle-cell disease with narcotic analgesics may have problematic value at best. To these difficulties on the side of the physician can be added the painful, disabling and unpredictable nature of the disease for the patient, which may lead to a variety of psychosocial difficulties. The interaction between these two groups of frustrated people -- physicians and patients -- in the hectic environment of the Emergency Department has the potential for considerable conflict.

It is against this backdrop that the present study of the physician-patient relationship at Yale-New Haven Hospital takes place. In formulating questions to answer in the present study, the literature review will be an important quide. Among others, questions suggested by the literature review include the following:

1) In light of the effects which age and sex appear to have on psychosocial adjustment in sickle-cell patients, how does hospital use at Yale-New Haven Hospital differ among sickle-cell patients of various ages, and between males and females?

2) In light of research showing positive effects of strategies such as management protocols on the care received by very frequent ED visitors, how many sickle-cell patients visit the YNHH Emergency Department with high frequency? How do their illness and their patterns of hospital use differ from those of less frequent ED visitors?

3) In light of research showing varying degrees of tension surrounding the issues of narcotic dependence and drug-seeking among patients with sickle-cell disease, how do issues of drug-seeking and drug abuse affect the care which patients receive at Yale-New Haven Hospital?

These questions have formed the basis of the design of the present study, and will be important in evaluating its results.

III. Methods

A. Methods -- General Introduction

The present study was conducted in three phases. In the first phase ("phase 1"), the goal was to characterize in general utilization of hospital services at Yale-New Haven Hospital by patients with sickle-cell disease. To this end, computer records of patient visits to YNHH and the YNHH ED over the period of one year (October 1, 1989 to October 1, 1990) were combed for visits by patients with sickle-cell disease, by reference to ICD-9 codes and complaint descriptions recorded for each visit. The frequency of ED visits and admissions could then be examined for various groups of patients, and breakdowns made by gender, age and frequency of ED visits. A particular goal of this process was to identify, if possible, a group of patients with sickle-cell disease with especially high utilization of ED services.

In the second phase of the study ("phase 2"), telephone interview was used to gain an understanding of the medical-care experiences of the group of high-frequency ED users. The interview in each case followed a standard format, and included both multiple-choice questions and open questions. Of the 12 patients with sickle-cell disease who used the ED with the greatest frequency during the study period, 4 could not be reached for various reasons. The full interview was done with 8 patients, including the top 6 ED users 19 years and older.

B. Phase 1

1. Phase 1 Data Collection

The methods of the first phase of the present study will be described first. A computer program was written to examine the billing records of YNHH over a one-year period (October 1, 1989 to October 1, 1990), in order to develope a general picture of the utilization of medical services by patients with sickle-cell disease.

The data examined in this phase of the study was originally contained in the general billing data of YNHH. The computer program designed to study this very large data base examined every "record" of the data base individually. Therefore, the first task will be to describe how a typical "record" is added to the data base, and what information it contains.

Each "record" of the data base represents data about an entire visit which is billable by YNHH (not only those bills which were actually collected). All the information about a single visit is kept under the same record. For example, every time a patient visits the YNHH ED, a record of the visit is generated, which includes basic demographic information (age, gender, address and phone number etc.), as well as some information about the visit itself (a short description of the complaint or diagnosis, eventual disposition, as well as the expenses incurred). After being seen in the ED, the patient might be discharged from the ED, and the record would then be closed. If, on the other hand, the patient goes on to be admitted, information on the admission (including discharge diagnoses, disposition, and, of course, expenses) is included under the same record. Thus, for a patient admitted to the hospital from the ED, registration information about the original ED visit is contained in the same record as information on the further admission to the hospital.

Patients who had incurred a hospital bill from any service of the hospital were identified and included in the patient sample. Expenses incurred by patients were classified by "source", that is, by the party that referred the patient for some test, treatment, or admission. The source might by the Emergency Department, a patient's private physician, the Yale Hematology Clinic (which functions as a quasi-independent practice group of Yale Faculty), or any of a number of institutions, such as nursing homes or Health Maintenance Organizations. For the purposes of the present study, the "source" of a given patient visit was classified to be either "Emergency Department" or "non-Emergency Department".

a. Operation of the Search Program: First Run

The computer program examined every visit record by all YNHH patients between October 1st, 1989 and October 1st, 1990. For each record encountered by the program, two fields of data were examined for clues that the visit was by a patient with sickle-cell disease, namely, the "complaint description" and the ICD-9 diagnosis codes. An indication of sickle-cell disease in either field led the program to copy the record to a data file. The search criteria for these two fields detail will now be described in greater detail.

The description of the patient's complaint, recorded at the patient's registration, was searched for an appearance of the words "sickle-cell crisis", "sickle crisis", "SSD crisis", or any of a number of combinations of such key words (see appendix for the actual search criteria). It should be noted that for a number of visits no complaint description was recorded.

The program also searched the ICD-9 codes recorded for each visit for the code "282.6" ([125]), indicating either sickle-cell disease or Sickle-C disease (ie double heterozygosity for hemoglobin S and hemoglobin C). Up to nine ICD-9 codes are assigned to each visit by specially trained coders in the Office of Professional services of YNHH. It should be noted that the ICD-9 diagnosis numbers were not infrequently inaccurate, as seen, for example, in the coding of 282.5, meaning "sickle-cell trait", as a diagnosis for patients with known homozygous sickle-disease. Such records with miscoded ICD-9 diagnoses would typically be picked up in spite of the miscoding by their complaint descriptions.

The program thus picked up all visits with a complaint description containing the key words "sickle cell" (etc.) OR with an ICD-9 code indicating sickle-cell disease. Though this process picked up patients with sickle-cell disease with what we felt was maximum sensitivity, it also picked up a number of spurious entries, usually identified by the complaint description. For example, the complaint description for one visit might be recorded as "SS pain crisis", which proved on inquiry to have referred to "substernal" pain crisis, or myocardial infarction. These spurious entries were usually obvious on manual inspection, and were removed.

Undoubtedly, other spurious entries remained undetected in the data file. One example of a spurious entry which might remain in the data file would be a visit with no recorded complaint description, and an ICD-9 code mistakenly recorded as indicating sickle-cell disease. In particular,

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[125] Note that the ICD-9 code "282.6" excludes sickle-cell thalasemia and sickle-cell trait. 282.6 does include the subcodes
282.60 ("sickle-cell anemia, uspecified"),
282.61 ("Hb-S disease without mention of crisis"),
282.62 ("Hb-S disease with mention of crisis"), and
282.63 ("Sickle-cell/Hb-C disease").
However, these subcategories were not completely reliably coded by the Office of Professional Services, so no further use was made of them.

patients with sickle-cell trait might conceivably be expecially likely to mistakenly receive an ICD-9 code for sickle-cell disease. The number of spurious entries is difficult to estimate, but it appears to be very small, judging from comparisons of the data file with a chart review of a limited number of patients (ie those chosen later for interview). However, time did not permit confirmation of the validity of the 1033 records by chart review.

b. Operation of the search program: second run

There remained an additional source of possible error in the data, namely, the fact that visits lacking a complaint description (or having, for example, a misspelling in a key word, such as "sikcle cell disease"), and also lacking a proper ICD-9 coding for sickle-cell disease, would escape detection. We could overcome this problem at least for patients who had been picked up on some other visit, by doing a second run through the data files.

In this second run, a second computer program (a modification of the first) searched through all records for the study period. This time, however, the program searched for all the patients (by unit number) who had been identified on the first run. This second run ensured that if a patient had been identified even once throughout the study period, all of his or her visits to YNHH in that period would be included in the final data file. Thus, the only visits that escaped detection were by patients who had been not been identified during any visit during the entire study period.

It is important to remember that this process of data collection aims to identify ED visits and hospital admissions by all patients with sickle-cell disease in the study period, not just incidents of sickle-cell crisis. Although a large proportion of the visits are likely to be for treatment of sickle-cell crisis pain, other visits may be for treatment of any number of other illness, which may or may not be related to sickle-cell disease. Unfortunately, neither the complaint descriptions for ED visits nor ICD-9 diagnosis codes were recorded accurately enough in hospital billing records for them to be useful to us in distinguishing crisis pain from other reasons to seek medical attention.

Another limitation to the process of data collection used here is an unavoidable selection bias towards patients who seek medical attention for their disease at YNHH. The data does not include patients in the community with mild disease, who passed the study year without seeking a doctor. Other patients may have seen a private physician during the year, but may not have been hospitalized at or been billed for services at YNHH. Still other patients may have preferred to travel to other hospitals to be treated for crisis pain. Because such patients are not included in the data, the data does not represent a sample of all the patients in the community with sickle-cell disease.

2. "Directly Counted" Variables

The original data file generated as described above contained 1033 records, each one corresponding to a visit to YNHH by a patient with sickle-cell disease between Oct. 1, 1989 and Oct. 1, 1990. We then counted the visits made by each patient, using a computer program specially written in dBase IV programming language. For each patient, the program counted

1) total visits
2) visits to the Emergency Department
3) total admissions
4) admissions via the Emergency Department
5) days spent in the hospital during admissions

In addition, a separate program counted Emergency Department visits and admissions for each month of the study period.

The data files generated by these counting programs -- now with each record corresponding to a single patient -- were then imported into EXCELL spread sheet format for analysis.

3. Patient ages

Age was calculated for each patient by subtracting the patient's birth year from 1990, which was the year into which most of the study period fell (Oct. 1, 1989 to Oct. 1, 1990).

For the purposes of the analysis, a "younger patient" was defined to be 18 years old of age or younger (ie born in 1972 or later), and an "adult" was defined to be 19 years of age or older (ie born 1971 or earlier). This

division of patients into "younger" and "older" patients does not necessarily reflect whether a patient was treated as a child or as an adult (ie in the pediatric vs. adult side of the ED, clinics, or inpatient units).

Three children with sickle hemoglobin were born during the study period, and stayed overnight, thereby accounting for three admissions. Sickle-cell disease was an incidental finding in these cases (found on routine screening), and not the reason for admission. These three children were removed from the data set.

C. Phase 2 Methods (interviews with patients)

1. The patient sample

Interviews with patients were at the heart of the present study. The group of patients targetted for interview were those sickle-cell patients 19 years and older with the highest utilization of emergency services at Yale-New Haven hospital, as identified by the computer survey of hospital visits by sickle-cell patients in the year-long period between 1 Oct. 1989 and 1 Oct. 1990 (phase 1). Approval by Human Investigations Committee was obtained before research commenced.

Originally, the 12 patients with the highest number of ED visits in the study period were chosen to be interviewed. All of them were sent a letter describing the study, and requesting their participation. Of the 12 patients, 4 could not be contacted: 1 Patient had an unlisted phone number, and did not visit Hematology Clinic or respond to letters. Another patient was found to have expired by gunshot in late 1990. Another patient was dropped from the list when letters to his address were returned, and no telephone number for the patient could be found. Still another patient agreed happily to be interviewed, but in spite of many interview appointments and phone calls was never at home with enough time to be interviewed.

The remaining 8 patients were interviewed in the time period from January 30, 1991 to March 7, 1991. Of these 8 patients, 2 patients had no telephone, but were able to be contacted when they visited Hematology Clinic. Interviews were then arranged with these 2 patients in person. The other 6 interviews were done by telephone.

The 8 patients interviewed included the 6 most frequent adult visitors to the ED with sickle-cell disease during the study year. The other 2

patients were ranked number 10 and number 12 in number of ED visits, respectively. The 8 patients together accounted for 179 Emergency Department visits during the study period, or 60% of all ED visits by adults (age > = 19) during the study period; numbers of ED visits among them ranged from 6 to 50. Three patients were male, and five were female.

The average age of the sample was 31.9 years, with ages ranging from 23 to 41. Two 23-year-old female patients, though in an age group normally seen by the adult medical service, continued to be seen by the pediatric service. Because the organization of services to these two patients was different than it was for patients seen on the adult side of the ED, they will be considered separately.

2. The interview format

Patients were interviewed by phone (or if necessary, in person), using a standard interview format. The format was based originally on the questions asked in the 1986 National Survey of Access to Health ([126]). Considerable changes were made in the original questions as they were adapted for use with sickle-cell disease patients. The survey questions are included in the appendix. The average interview duration for the 8 interviews was 57 minutes, with a range of 45 to 70 minutes.

Briefly, the survey asked questions in the following areas:

1) Where does the patient receive his or her care?

2) Experiences with Hematology Clinic

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[126] Freeman, 1986. Note that no attempt was made to compare the answers to questions in the present study to corresponding answers to the 1986 survey

3) Experiences with the Emergency Department

4) Experiences as an inpatient

5) Compliance with doctor's orders

6) Disability

7) Pain medication

8) Access to medical care

9) General Background, education, marital status

Many of the questions were multiple choice, to allow comparison of responses by different patients. However, for most of the the sections there was an open question to record each patient's responses in his or her own words.

IV. Results

A. Phase 1 Results

1. Introduction to phase 1 results

In this section, results of the computer survey of visits of patients with Sickle-cell disease to Yale-New Haven from Oct.1,1989 to Oct.1,1990 will be presented. The results will be organized to answer the following questions:

1) Who were the the patients with sickle-cell disease seen at Yale-New Haven Hospital? What what was their overall distribution in terms of age and gender?

2) How much did sickle-cell patients use hospital services in toto?

3) How did hospital use differ for patients of different age groups and between males and females?

4) How did hospital use differ for patients with various levels of utilization of ED services? In particular, what patterns of hospital use can be found among patients with sickle-cell disease who frequently visit the Emergency Department?

5) Do visits to the Emergency Department follow cyclical patterns over time? Especially, does the frequency of visits vary with temperature or season? If so, are these patterns different for young patients than for older patients?

These questions will be answered by reference to 10 charts, which will be provided in the succeeding pages. For more exact numbers, the reader

may turn to the 7 tables of the results. placed next to the corresponding charts.

2. An overview of the sample of patients with sickle-cell disease

See tables 1 and 2, and charts 1 and 2

A total of 132 patients with sickle-cell disease were identified. They had a mean age of 21 years. 48 were male. and 84 were female.

Patients visited the Emergency Department a total of 422 times during the study year. This represented an average of 3.2 visits per patient per year. Patients in the sample were admitted a total of 164 times. and they spent a total of 969 days admitted in the hospital. Admissions lasted a mean average of 5.9 days long.

Of the 422 visits to the Emergency Department. 131 (31.0%) resulted in admission. Of the 164 admissions. 131 (79.9%) were via the Emergency Department.

3. Analysis of results by gender and age

See tables 1 through 4, and charts 1 through 4

Chart number 1 shows a breakdown of hospital use by age groups. As these data demonstrate, sickle-cell patients seen at Yale are predominantly young. Of the 132 patients, 33 (25%) were 10 years or younger, and 33 (25%) were between 11 and 20. Thus, of all the patients seen, 50% were in the first two decades of life.

The 21-to-30 age group was the largest, with 35 patients (27% of the total number). But in age groups older than this, numbers fell off sharply. Only 21 patients (15% of the total number) were in the 31-to-40 year group, and 6 patients (5% of total) were in the 41-50 age group. Only 4 patients in the sample were older than 51 years.

Visits to the Emergency Department varied widely among different age groups. By far the greatest proportion of ED visits was accounted for by members of the 21-30 year age group. Patients between 21 and 30 years accounted for 37.9% of all ED visits, out of proportion to their numbers (26.5% of total) ([127]). As graph 1 shows, patients in successive age groups to "either side" of the 21-30 year age group visited the ED proportionately less and less.

Chart 1 demonstrates a similar peak in the numbers of admissions and hospital days for various age groups; this time, however, the peak lies in the 11-20-year age group. Patients in this group were admitted 47 times (32% of

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[127] It is of note that the 3 patients who visited the ER with the greatest frequency were all over 30 years old. Thus, the peak of ER visits in this age group is not simply due to skewing by the 3 top ER visitors.

total) and spent 312 days in the hospital (32% of total), somewhat out of proportion to their numbers (25% of total).

There was a striking predominance of females in the sample (See charts 2,3 and 4). Of 132 patients in the total sample, 84 (64%) were female, and 48 (36%) were male. These gender proportions were similar both for patients 18 years and younger and for patients 19 years and older.

Use of emergency services by patients with sickle-cell disease at Yale-New Haven Hospital differed somewhat by gender. As chart 2 shows, female patients, partly reflecting their greater numbers in the sample, predominated overall in total visits to the ED, with 56% of ED visits by females, and 43.1% by men. However, actual "per patient" visits to the ED were slightly higher for men (3.8 ED visits/patient) than for women (2.9 ED visits/pt).

Among patients 18 years and younger (table/chart 3), the predominance of total ED visits by females was much more striking, with 84% of ED visits by females, and 16% by males. Among these younger patients, "per person" visits to the ED were higher for women (2.5 ED visits per patient) than for men (1.0 ED visits per patient).

Among all patients 19 years or older (table/chart 4), however, the female dominance of ED use was reversed, with males accounting for 54.6% of ED visits, and females for 45.4%. On a "per person" basis, the gender difference was even more striking, with 5.8 ED visits per male patient, versus 3.2 ED visits per patient for females.

Care must be taken in assessing the importance of this male predominance in ED use by sickle-cell patients 19 years and older. In fact, this male predominance appears on closer examination to be at least partially due to a skewing effect by a very small group of patients with very high frequency of ED use. When, for example, the three patients with the highest

frequency of ED use (two males and a female, accounting together for 119 ED visits) were temporarily removed from the data set, female predominance in use of the ED reappeared (see chart 4a).

In numbers of admissions and hospital days for all patients, female patients predominated as well, again at least partially reflecting their larger numbers in the sample (see table/chart 2). Female patients overall accounted for 72.6% of all admissions for sickle-cell patients, and 72.5% of hospital days, versus 27.4% of admissions and 27.6% of hospital days for males. Morover, on a "per patient" basis overall, female patients were admitted more frequently (1.42 admits/patient) than males (.94 admits/patient). However, average length of hospital stay was similar overall for men (5.9 days/admission) and women (5.9 days/admission).

In summary, the sample of sickle-cell patients included 132 patients with a mean age of 21 years. There was a striking predominance of young patients, with 50% of patients under 20 years of age, and only 4 patients over the age of 50 years. Of all hospital use by members of the sample, the largest proportion of ED visits was accounted for by patients in the 21-30-year age group, and the largest proportion of admissions and hospital days was accounted for by patients in the 11-20-year age group. In addition, there was a striking predominance of female patients in the sample, with females making up 64% of the sample overall. Female patients overall accounted for 57% of ED visits, 73% of total admissions, and 72% of total hospital days. The female predominance in patient numbers and in hospital use was greater in younger patients than in older patients.

4. Analysis of results by number of ED visits

See tables 5 and 6, and charts 5 and 6

The variation in frequency of use of emergency services among sickle-cell patients in the sample was very wide. It was observed at the outset that a small number of patients had visited the ED with very high frequency. For example, the five adult patients with the highest ED use had visited the ED 50 times, 37 times, 32 times, 22 times, and 15 times respectively during the study year. Many other patients had visited only once or twice during the study year, or not at all.

In order to study overall hospital use by patients with different levels of use of emergency services, the sample of sickle-cell patients (132 patients) was stratified into four groups based on number of ED visits during the study year. The groups were arbitrarily defined as follows:

Group 1: No ED visits at all
Group 2: Between 1 and 5 ED visits
Group 3: Between 6 and 10 ED visits
Group 4 11 or more ED visits

Data were analyzed separately for younger patients and adult patients. It is important to note that this division, as well, is based on an arbitrary choice, namely, on the choice of 18/19 years of age as the dividing point. This division is convenient for the sake of analysis, and offers a simple way to ascertain whether the patterns of hospital use are grossly different for children and for adults with sickle-cell disease. However, this division may not, for example, reflect whether patients in each group are seen on the

pediatric or on the adult side of the ED. As will be seen, a number of patients in their early 20's continued to be cared for by the pediatric service.

The following chart summarizes data on ED visits (for more detail see tables 5 and 6):

Visits to the Emergency Department by Patients at Yale-New Haven Hospital from October 1, 1989 to October 1,1990:

	Younger Patients (age <= 18 yr)			Adult Patients (age >= 19 yr)		
	Pt. #	ED visits #	(%)	Pt. #	ED visits #	(%)
Group 1 (no ED visits)	23	0	(0%)	19	0	(0%)
Group 2 (1-5 ED visits)	33	62	(50%)	38	81	(27%)
Group 3 (6-10 ED visits)	4	30	(24%)	8	60	(20%)
Group 4 (11+ ED visits)	2	33	(26%)	5	156	(53%)
TOTALS	62	125	(100%)	70	297	(100%)

a. Younger patients (18 years and younger)

Among the 62 sickle-cell patients who were 18 or younger (see table/chart 5) during the study year, the majority (33 patients) had between 1 and 5 ED visits, 4 patients had between 6 and 10 visits, and only 2 patients had 11 or more visits. The greatest number of visits in this group was by a 2-year old female patient, who visited the ED 21 times during the study year.

Younger patients in groups and 3 and 4, as was expected from the group selection criteria, accounted for ED visits out of proportion to their number. The 6 patients in these two groups (about 10% of the total group of younger patients) accounted together for 63 visits (about 50% of all ED visits by younger patients).

Younger patients who visited the ED frequently were also hospitalized more often, and stayed in the hospital a greater number of total days, than patients who visited the ED less frequently. Patients in groups 3 and 4 were admitted an average of 4.8 and 6.5 times per patient, respectively, compared with .3 times per patient among patients in group 1, and 1.2 times per patient in group 2. However, average length of hospital stay (as shown in table 5, left-most column) was relatively similar for all groups, with an average of 5.1 days per admission.

b. Adult Patients (19 years and older)

Among the 70 sickle-cell patients who were 19 or older (see table/chart 6) during the study year, 19 patients did not visit the ED at all, 38 patients visited between 1 and 5 times, 8 visited between 6 and 10 times, and 5 patients visited 11 or more times. The greatest number of visits by an adult was by a 30-year old male, who visited the ED 50 times during the study year.

Adult patients in groups 3 and 4 (ie with 6 or more ED visits) visited the ED far out of proportion to their numbers. The 13 patients in these two groups (representing about 19% of adults in the sample) accounted for 216 ED visits, or about 73% of all ED visits by adults. In fact, the 5 patients in group 4 alone, though representing only 7% of adult patients, accounted for about 53% of all ED visits by adults.

As with younger patients, adult patients who visited the ED more frequently were also hospitalized more often, and stayed in the hospital a greater number of days, than patients who visited the ED less often. Adult patients in groups 3 and 4 were admitted an average of 3.3 and 3.4 times per patient, respectively, compared with .4 times per patient among patients in group 1, and .9 times per patient among patients in group 2.

Average length of hospital stay for all adult sickle-cell patients (as shown in table 6, left-most column) was 6.6 days per admission. Patients in group 1 (ie patients who had not visited the ED, and thus who were not admitted via the ED) had an especially high average length of hospital stay, at 10.5 days per admission. Conversely, patients in group 4 (ie those with at least 11 visits) had a relatively shorter length of hospital stay, at 4.8 days per admission.

c. Summary of data on frequent ED visitors

In summary, the most striking common theme among both younger patients and adults was the disproportionate number of total ED visits accounted for by small groups of very frequent ED visitors. Among patients in groups 3 and 4 (ie those with 6 or more ED visits), 6 younger patients (10% of all younger patients) accounted for 50% of all ED visits by younger patients, whereas 13 adult patients (19% of all adult patients) accounted for 73% of ED visits by adults.

Note that this predominance of ED use by a small group of frequent ED users was much more marked in adults than in younger patients. Indeed, among adult patients, those patients with 6 or more ED visits were greater in number (13 adult patients vs. 6 younger patients) and accounted for a higher proportion of total ED use (73% of visits by adults vs. 50% of visits by younger patients).

Among both younger patients and adults, patients who visited the ED frequently were also admitted more frequently on average, and spent more days in the hospital, than patients who visited the ED less frequently.

5. Analysis of results by month

See table 7, and charts 7 through 10

It has been often reported that sickle-cell crisis may be evoked in many patients by cold weather, or by rapid changes in temperature. One might thus expect that during cold weather that visits to the ED by sickle-cell patients

would increase, or that patients would be admitted more frequently. Therefore, it was of interest to investigate seasonal variations in hospital use.

To this end, numbers of ED visits and admissions were calculated for each month of the study period, for the group as a whole, as well as for younger patients (18 years and younger) and for older patients (19 years and older). These were plotted against average daily temperatures for each month, as reported by the United Illuminating Company, a local electric utility company. The results may be found in table 7 and charts 7 through 10.

The number of Emergency Department visits and admissions varied widely by month. Monthly ED visits during the study period ranged from 19 to 47 visits, with a mean average of 35.2 ED visits/month. Monthly admissions ranged from 7 to 20, with a mean average of 13.7 admissions/month.

In order to evaluate these monthly fluctuations in Emergency Department visits and admissions, numbers of ED visits and admissions were totalled for the four months during the study period when average daily temperatures were under 40 degrees F (December, January, February, and March), and also for the four months when average daily temperatures were above 60 degrees F (June, July, August and September).

Put briefly, no increases in Emergency Department visits were found during colder months of the 1-year study period. As can be seen in table 7, numbers of ED visits and admissions were very similar for cold and warm months. For all patients, in fact, ED visits during the four "cold" months (142 ED visits) were actually slightly fewer than in the four "warm" months (150 ED visits). Numbers of admissions for all patients were also similar during "cold" months (60 admissions) and "warm" months (58 admissions).

Neither was a seasonal variation in hospital use seen among younger patients as group (see table 7, chart 8), nor adult patients as a group (table 7, chart 9). Numbers of Emergency Department visits and admissions were similar during warm and cold months for both younger and adult patients. The only case in which a cyclical pattern appears to exist, as seen visually in a rising and falling curve, is in admissions for younger patients over the study year, with a nadir in a warm month (May) and a peak in a cold month (December) (see chart 9). But as remarked, no significant difference was found in younger patients' total admissions over the 4 "cold" and the 4 "warm" months.

It should be remarked here that the method followed here only roughly examines the effect of temperature on sickle crisis. Figures were totalled by month, and no attempt was made to quantify the increased risk of sickle crisis based on the temperature during any individual day, or any time of day.

In summary, there was wide variation in numbers of Emergency Department visit and admissions by month during the study period. However, no cyclical or seasonal pattern could be observed, neither in sickle-cell patients as a whole, nor in younger patients nor adult patients as groups. The present analysis indicates that total ED visits and admissions among sickle-cell patients did not increase during cold months.

6. Summary of computer survey results

Visits to YNHH by patients with sickle-cell disease, 1 Oct. 1898 to 1 Oct. 1990

The computer survey identified a total of 132 patients with sickle-cell disease, with a mean age of 21 years. Of the 422 visits to the Emergency Department accounted for by sickle-cell patients during the study year, 131 (31.0%) resulted in admission. These 131 ED admissions represented 79.9% of the 164 total admissions for the group. Admissions overall lasted an average of 5.9 days each.

Results of the computer survey were analyzed by age, gender, and frequency of ED visits, as well as by month. The following findings were especially striking:

a.) There was a striking predominance of young patients, with 50% of patients under 20 years of age, and only 4 patients over the age of 50 years.

b.) There was a striking predominance of female patients in the sample, with females making up 64% of the sample overall. Female patients overall accounted for 57% of ED visits, 73% of total admissions, and 72% of total hospital days. The female predominance in patient numbers and in hospital use was greater in younger patients than in older patients.

c.) The most striking common theme among both younger patients and adults was the disproportionate number of total ED visits accounted for by small groups of very frequent ED visitors. Among patients with 6 or more ED visits during the study year, 6 younger patients (10% of all younger patients) accounted for 50% of all ED visits by younger patients, whereas 13 adult patients (19% of all adult patients) accounted for 73% of ED visits by adults.

d.) Among both younger patients and adults, patients who visited the ED frequently were also admitted more frequently on average, and spent more days in the hospital, than patients who visited the ED less frequently.

e.) There was wide variation in numbers of Emergency Department visit and admissions by month during the study period. However, no cyclical or seasonal pattern could be observed, neither in sickle-cell patients as a whole, nor in younger patients or adult patients as groups.

7. Tables and charts -- phase 1

TABLE 1 (CORRESPONDS TO CHART 1) BREAKDOWN BY AGE GROUPS SICKLE CELL PATIENTS SEEN AT YALE NEW HAVEN HOSPITAL FROM OCT 1 1989 TO OCT 1 1990

Conditions for inclusion		PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT
AGE >=0	AGE <=10	33	25.00%	145	14.92%	(4.39)	55	13.03%	(1.67)	28	17.07%	(0.85)	24	18.32%	(0.73)	108	11.15%	(3.27)
AGE >=11	AGE <=20	33	25.00%	209	21.50%	(6.33)	72	17.06%	(2.18)	52	31.71%	(1.58)	38	29.01%	(1.15)	312	32.20%	(9.45)
AGE >=21	AGE <=30	35	26.52%	284	29.22%	(8.11)	160	37.91%	(4.57)	47	28.66%	(1.34)	38	29.01%	(1.09)	288	29.72%	(8.23)
AGE >=31	AGE <=40	21	15.91%	234	24.07%	(11.14)	87	20.62%	(4.14)	28	17.07%	(1.33)	22	16.79%	(1.05)	200	20.64%	(9.52)
AGE >=41	AGE <=50	6	4.55%	71	7.30%	(11.83)	45	10.66%	(7.50)	8	4.88%	(1.33)	8	6.11%	(1.33)	53	5.47%	(8.83)
AGE >=51	AGE <=60	3	2.27%	26	2.67%	(8.67)	3	0.71%	(1.00)	1	0.61%	(0.33)	1	0.76%	(0.33)	8	0.83%	(2.67)
AGE >=61	AGE <=70	1	0.76%	3	0.31%	(3.00)	0	0.00%	(0.00)	0	0.00%	(0.00)	0	0.00%	(0.00)	0	0.00%	(0.00)
TOTALS FOR ALL PTS		132	100.00%	972	100.00%	(7.36)	422	100.00%	(3.20)	164	100.00%	(1.24)	131	100.00%	(0.99)	969	100.00%	(7.34)

KEY

1) "Plain" numbers represent absolute numbers for each variable
2) Numbers followed by "%" represent percentages of the TOTAL of the corresponding variable
3) Numbers in parentheses "( )" represent ratios of each variable PER PATIENT

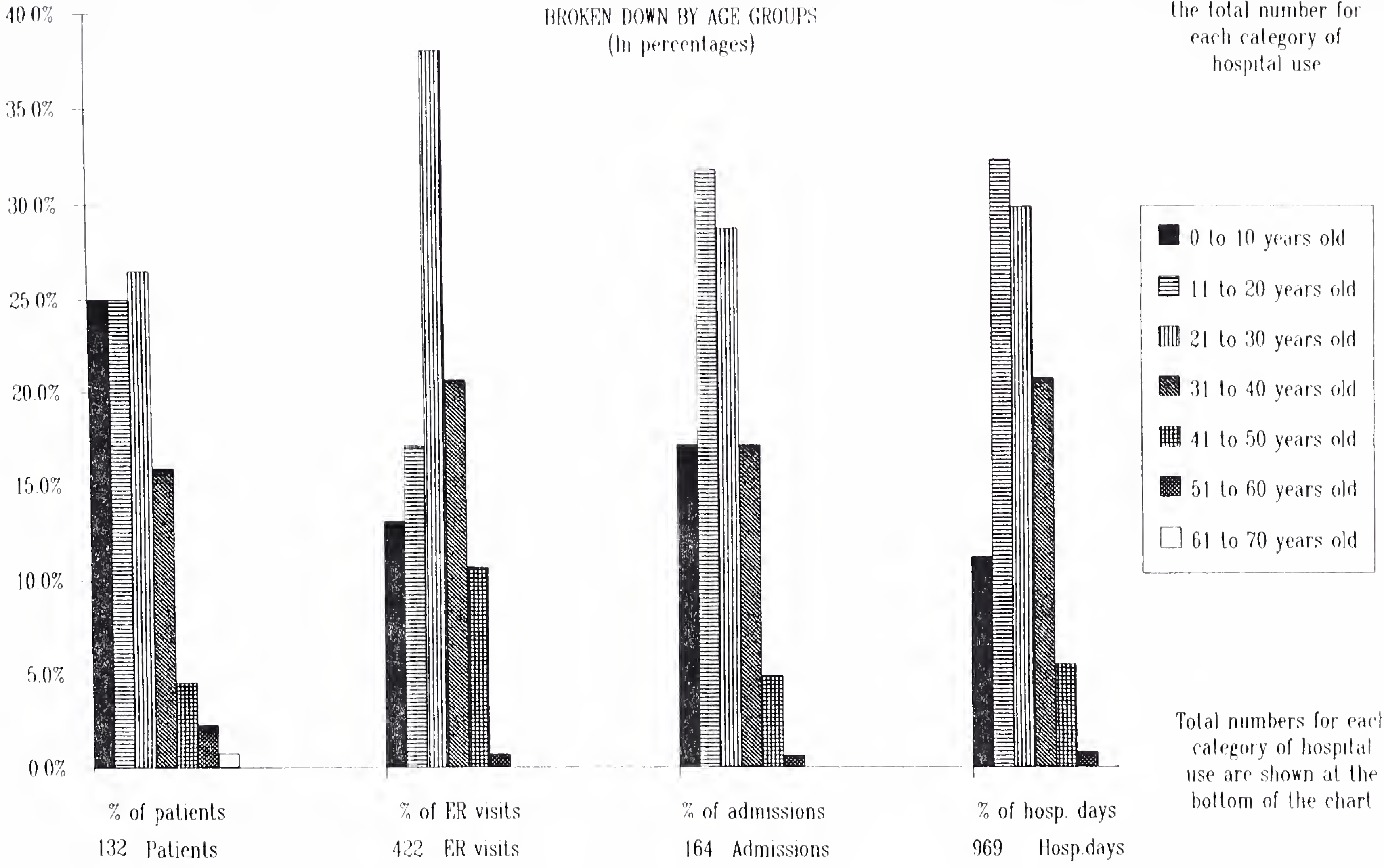
ER admits/total admits 79.9%

ER admits/ER visits 31.0%

Mean hosp days/admission 5.9

D T Fleming YMS

CHART NUMBER 1
HOSPITAL USE BY SICKLE CELL PATIENTS.
OCT.1, 1989 TO OCT 1,1990, AT YNHH
BROKEN DOWN BY AGE GROUPS
(In percentages)



All graphed numbers are shown as percentages of the total number for each category of hospital use

Total numbers for each category of hospital use are shown at the bottom of the chart

Douglas T Fleming, YMS

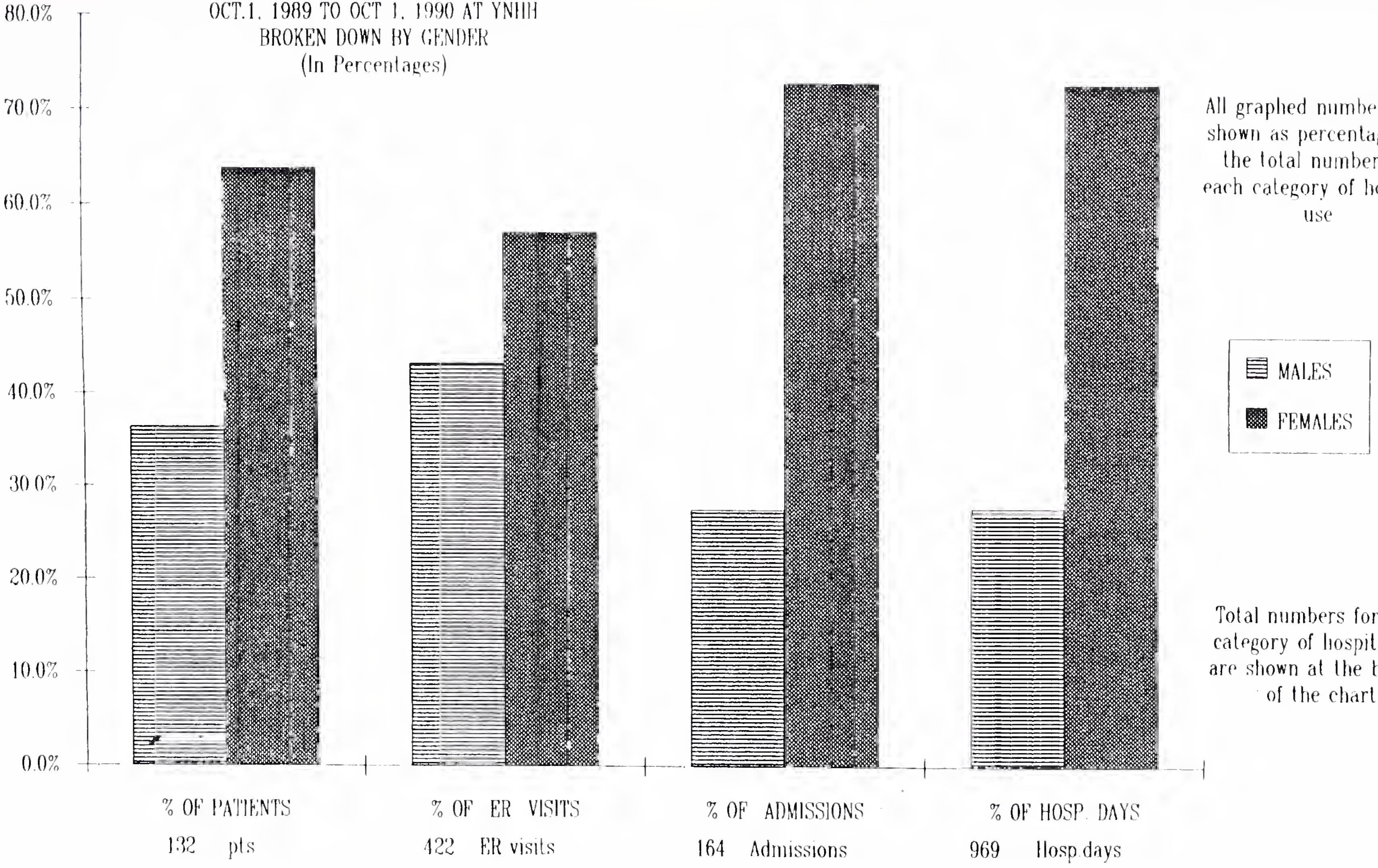
TABLE 2 (CORRESPONDS TO CHART 2)

BREAKDOWN BY GENDER -- SICKLE-CELL PATIENTS SEEN AT YALE NEW HAVEN HOSPITAL FROM OCT 1 1989 TO OCT 1 1990
INCLUDES PATIENTS OF ALL AGES

Conditions for inclusion			PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT
AGE >=0	TOT_VIS >=0	SEX M	48	36.36%	360	37.04%	(7.50)	182	43.13%	(3.79)	45	27.44%	(0.94)	36	27.48%	(0.75)	267	27.55%	(5.56)
AGE >=0	TOT_VIS >=0	SEX F	84	63.64%	612	62.96%	(7.29)	240	56.87%	(2.86)	119	72.56%	(1.42)	95	72.52%	(1.13)	702	72.45%	(8.36)
		TOTALS:	132	100.00%	972	100.00%	(7.36)	422	100.00%	(3.20)	164	100.00%	(1.24)	131	100.00%	(0.99)	969	100.00%	(7.34)

D T Fleming YMS

CHART NUMBER 2
HOSPITAL USE BY ALL SICKLE CELL PATIENTS
OCT.1, 1989 TO OCT 1, 1990 AT YNHH
BROKEN DOWN BY GENDER
(In Percentages)



All graphed numbers are shown as percentages of the total number for each category of hospital use

Total numbers for each category of hospital use are shown at the botton of the chart

Douglas T Fleming, YMS

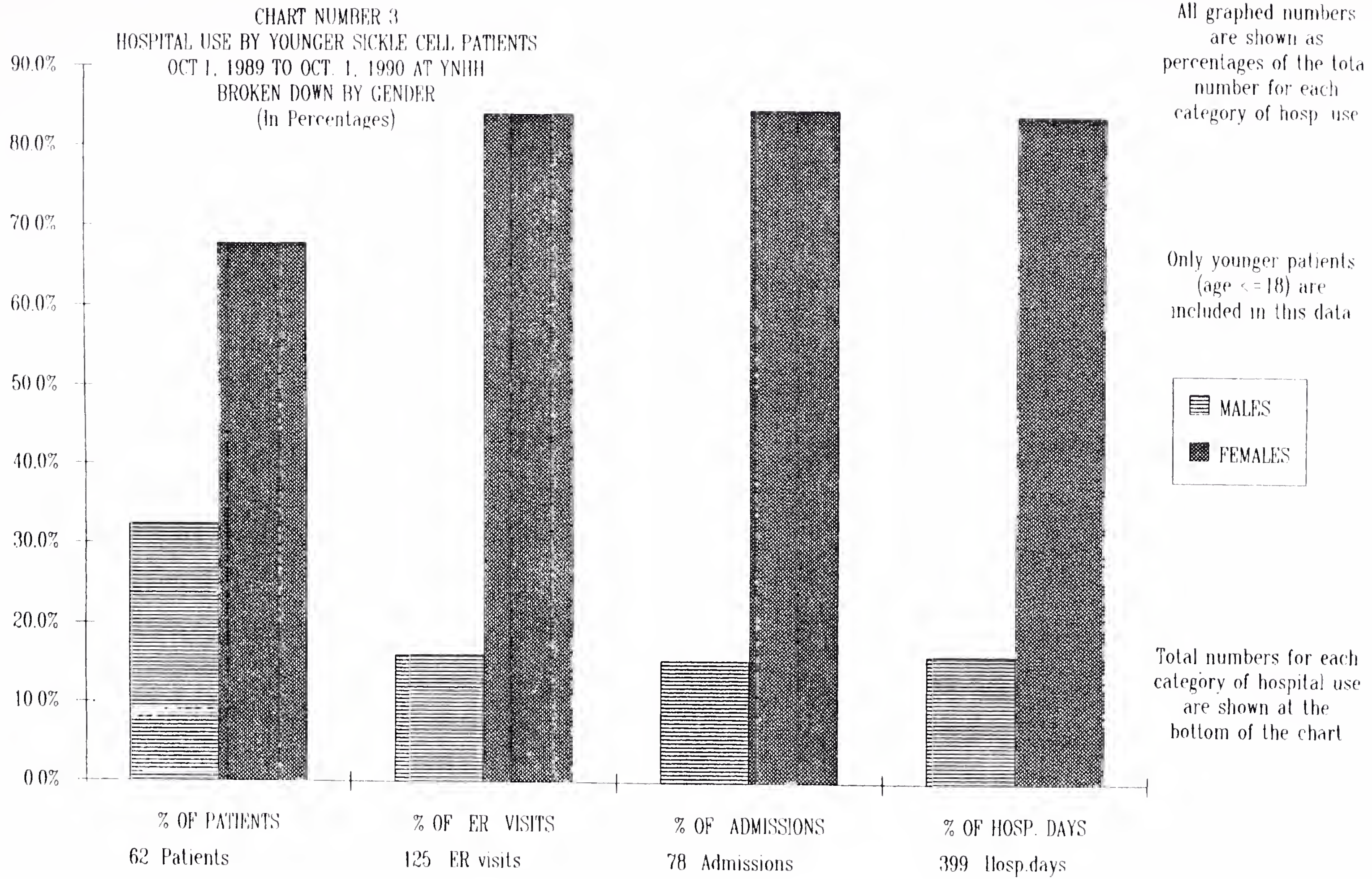
TABLE 3 (CORRESPONDS TO CHART 3)

BREAKDOWN BY GENDER - SICKLE CELL PATIENTS SEEN AT YALE NEW HAVEN HOSPITAL FROM OCT 1.1989 TO OCT 1 1990
INCLUDES ONLY PATIENTS 18 YEARS AND YOUNGER

Conditions for inclusion			PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT
AGE <=18	TOT_VIS >=0	SEX M	20	32.26%	85	24.85%	(4.25)	20	16.00%	(1.00)	12	15.38%	(0.60)	9	14.75%	(0.45)	64	16.04%	(3.20)
AGE <=18	TOT_VIS >=0	SEX F	42	67.74%	257	75.15%	(6.12)	105	84.00%	(2.50)	66	84.62%	(1.57)	52	85.25%	(1.24)	335	83.96%	(7.98)
		TOTALS	62	100.00%	342	100.00%	(5.52)	125	100.00%	(2.02)	78	100.00%	(1.26)	61	100.00%	(0.98)	399	100.00%	(6.44)

D T Fleming YMS

CHART NUMBER 3
HOSPITAL USE BY YOUNGER SICKLE CELL PATIENTS
OCT 1, 1989 TO OCT. 1, 1990 AT YNHH
BROKEN DOWN BY GENDER
(In Percentages)



All graphed numbers
are shown as
percentages of the tota
number for each
category of hosp use

Only younger patients
(age <=18) are
included in this data

Total numbers for each
category of hospital use
are shown at the
bottom of the chart

Douglas T. Fleming, YMS

TABLE 4 (CORRESPONDS TO CHART 4)

BREAKDOWN BY GENDER -- SICKLE CELL PATIENTS SEEN AT YALE-NEW HAVEN HOSPITAL FROM OCT 1.1989 TO OCT 1.1990
INCLUDES ONLY PATIENTS 19 YEARS AND OLDER

Conditions for inclusion

AGE	TOT_VIS	SEX	PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT.	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT
>=19	>=0	M	28	40.00%	275	43.65%	(9.82)	162	54.55%	(5.79)	33	38.37%	(1.18)	27	38.57%	(0.96)	203	35.61%	(7.25)
>=19	>=0	F	42	60.00%	355	56.35%	(8.45)	135	45.45%	(3.21)	53	61.63%	(1.26)	43	61.43%	(1.02)	367	64.39%	(8.74)
		TOTALS	70	100.00%	630	100.00%	(9.00)	297	100.00%	(4.24)	86	100.00%	(1.23)	70	100.00%	(1.00)	570	100.00%	(8.14)

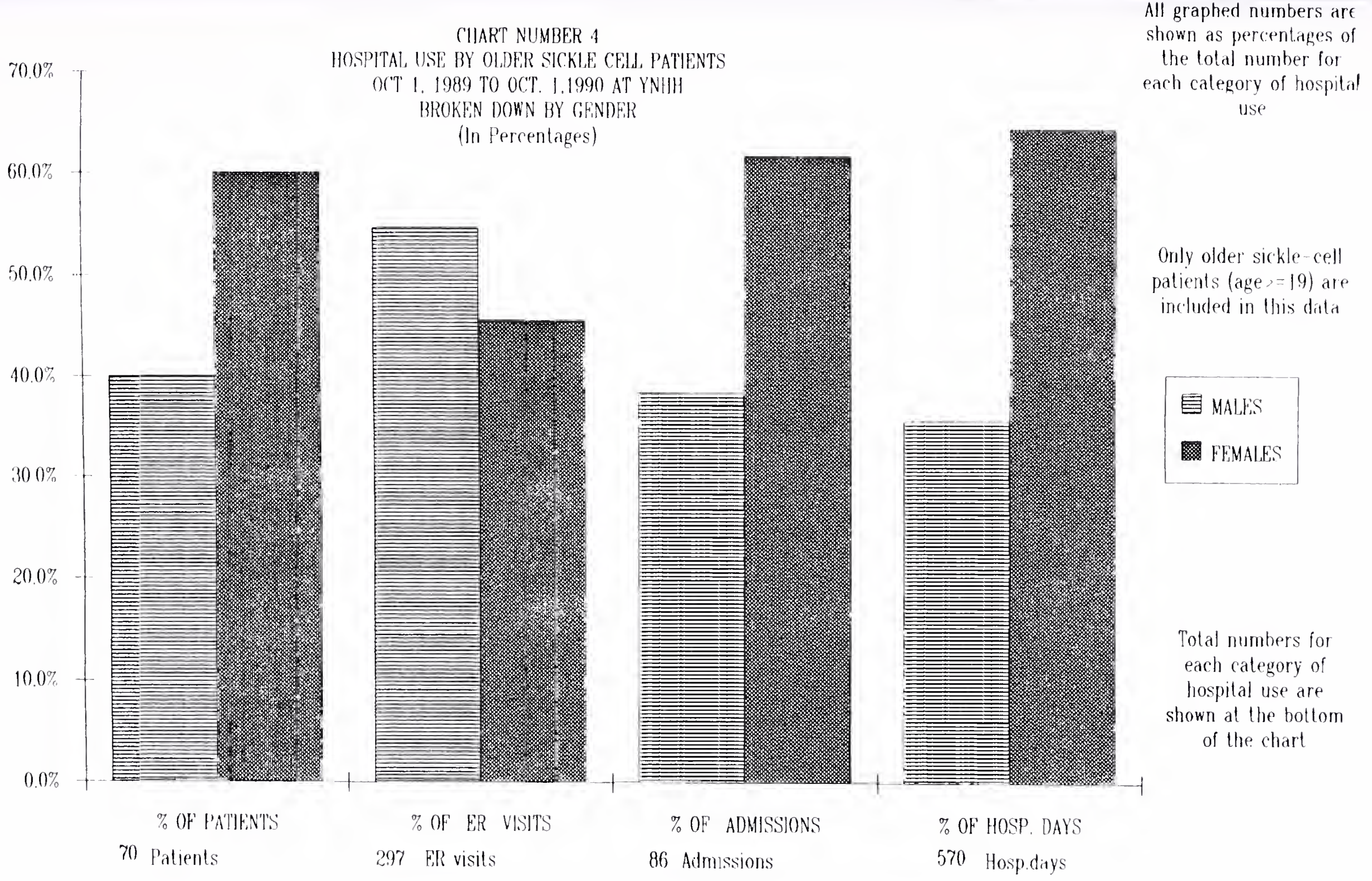


CHART NUMBER 4
HOSPITAL USE BY OLDER SICKLE CELL PATIENTS
OCT 1, 1989 TO OCT. 1,1990 AT YNHH
BROKEN DOWN BY GENDER
(In Percentages)

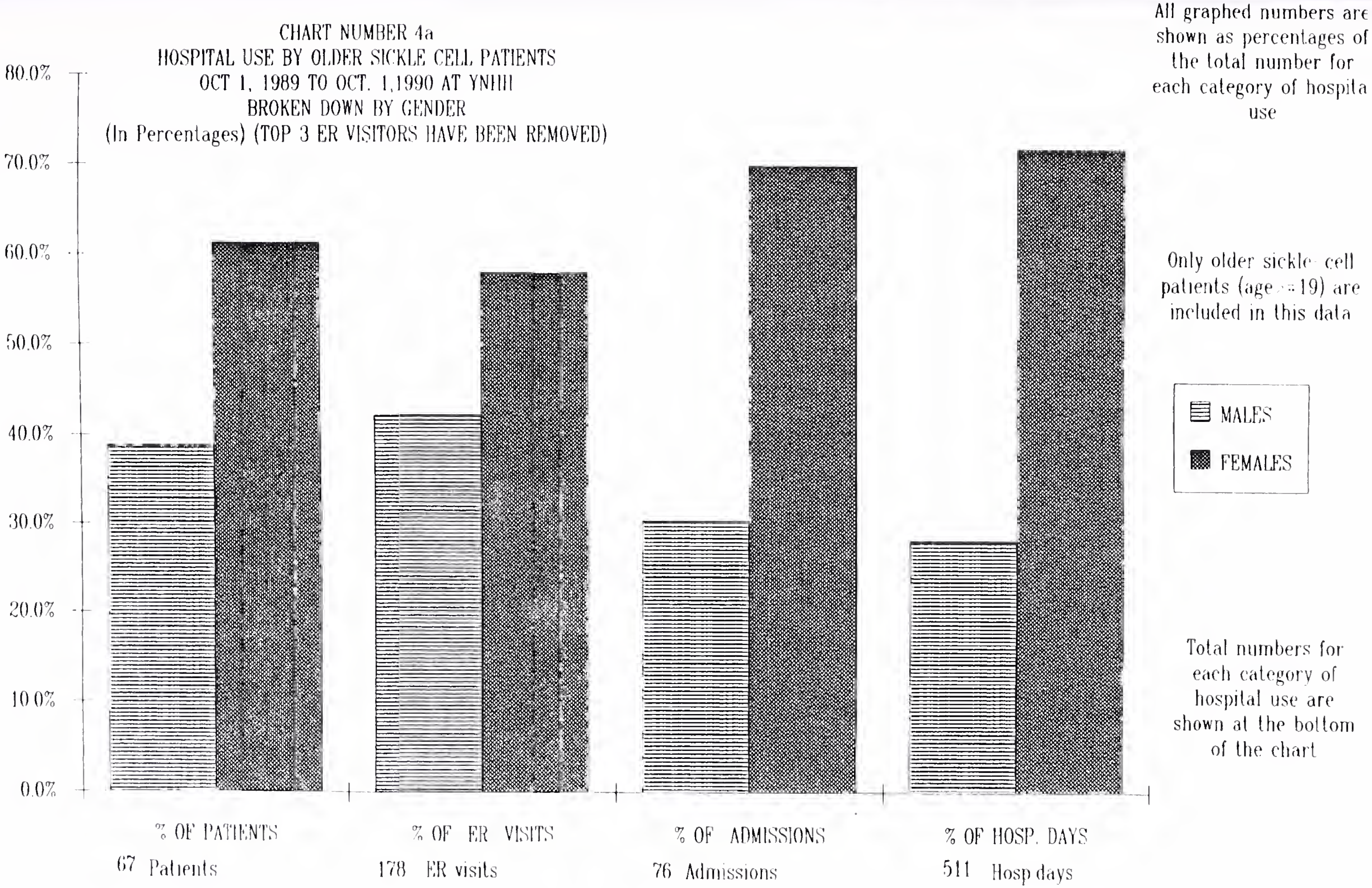
All graphed numbers are shown as percentages of the total number for each category of hospital use

Only older sickle-cell patients (age >=19) are included in this data

Total numbers for each category of hospital use are shown at the bottom of the chart

Douglas T Fleming, YMS

CHART NUMBER 4a
HOSPITAL USE BY OLDER SICKLE CELL PATIENTS
OCT 1, 1989 TO OCT. 1,1990 AT YNHH
BROKEN DOWN BY GENDER
(In Percentages) (TOP 3 ER VISITORS HAVE BEEN REMOVED)



All graphed numbers are shown as percentages of the total number for each category of hospita use

Only older sickle cell patients (age >= 19) are included in this data

Total numbers for each category of hospital use are shown at the bottom of the chart

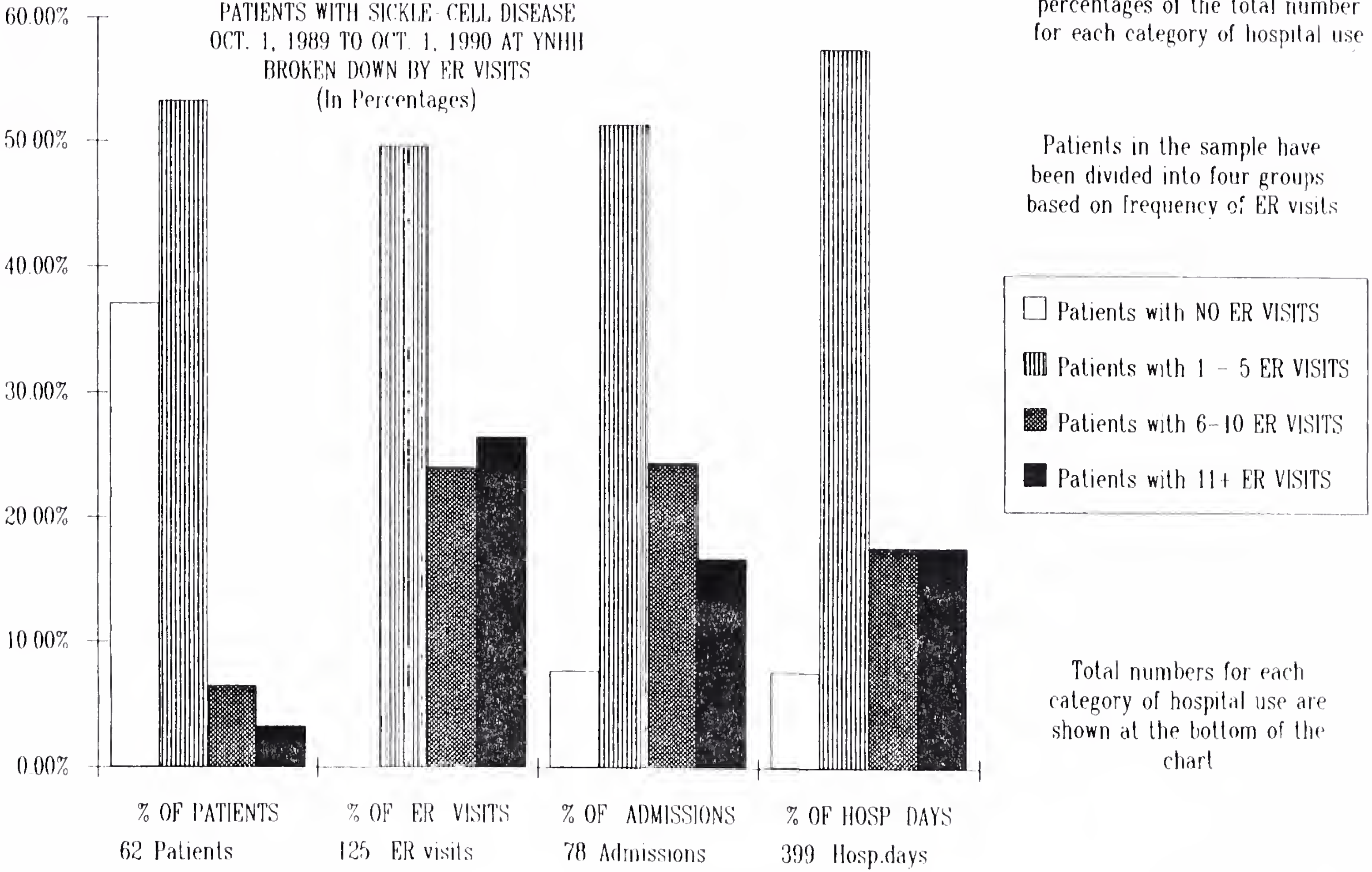
Douglas T. Fleming, YMS

TABLE 5 (CORRESPONDS TO CHART 5)

BREAKDOWN BY ER VISITS - - SICKLE-CELL PATIENTS SEEN AT YALE NEW HAVEN HOSPITAL FROM OCT 1,1989 TO OCT 1,1990
INCLUDES ONLY PATIENTS 18 AND YOUNGER

Conditions for inclusion			PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP. DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT	DAYS PER ADMIT
AGE	ER_VIS	ER_VIS																		
<=18	0	0	23	37.10%	81	23.68%	(3.52)	0	0.00%	(0.00)	6	7.69%	(0.26)	0	0.00%	(0.00)	30	7.52%	(1.30)	5.0
AGE	ER_VIS	ER_VIS																		
<=18	>=1	<=5	33	53.23%	157	45.91%	(4.76)	62	49.60%	(1.88)	40	51.28%	(1.21)	33	54.10%	(1.00)	229	57.39%	(6.94)	5.7
AGE	ER_VIS	ER_VIS																		
<=18	>=6	<=10	4	6.45%	60	17.54%	(15.00)	30	24.00%	(7.50)	19	24.36%	(4.75)	17	27.87%	(4.25)	70	17.54%	(17.50)	3.7
AGE	ER_VIS	ER_VIS																		
<=18	>=11	<60	2	3.23%	44	12.87%	(22.00)	33	26.40%	(16.50)	13	16.67%	(6.50)	11	18.03%	(5.50)	70	17.54%	(35.00)	5.4
	TOTALS		62	100.00%	342	100.00%	(5.52)	125	100.00%	(2.02)	78	100.00%	(1.26)	61	100.00%	(0.98)	399	100.00%	(6.44)	5.1

CHART NUMBER 5
HOSPITAL USE BY YOUNGER (AGE <=18)
PATIENTS WITH SICKLE-CELL DISEASE
OCT. 1, 1989 TO OCT. 1, 1990 AT YNHH
BROKEN DOWN BY ER VISITS
(In Percentages)



All graphed numbers are shown as percentages of the total number for each category of hospital use

Patients in the sample have been divided into four groups based on frequency of ER visits

Total numbers for each category of hospital use are shown at the bottom of the chart

Douglas T Fleming, YMS

TABLE 6 (CORRESPONDS TO CHART 6)

BREAKDOWN BY ER VISITS -- SICKLE-CELL PATIENTS SEEN AT YALE NEW HAVEN HOSPITAL FROM OCT 1,1989 TO OCT 1,1990
INCLUDES ONLY PATIENTS 19 YEARS AND OLDER

Conditions for inclusion			PATIENT NUMBER	PATIENT NUMBER % OF TOT	TOTAL VISITS	TOTAL VISITS % OF TOT	TOTAL VISITS PER PT	ER VISITS	ER VISITS % OF TOT	ER VISITS PER PT	TOTAL ADMITS	TOTAL ADMITS % OF TOT	TOTAL ADMITS PER PT	ER ADMITS	ER ADMITS % OF TOT	ER ADMITS PER PT	HOSP DAYS	HOSP DAYS % OF TOT	HOSP DAYS PER PT	DAYS PER ADMIT
AGE	ER_VIS	ER_VIS																		
>=19	0	0	19	27 14%	99	15 71%	(5 21)	0	0 00%	(0 00)	8	9 30%	(0 42)	0	0 00%	(0 00)	84	14 74%	(4 42)	10 5
AGE	ER_VIS	ER_VIS																		
>=19	>=1	<=5	38	54 29%	229	36 35%	(6 03)	81	27 27%	(2 13)	35	40 70%	(0 92)	28	40 00%	(0 74)	242	42 46%	(6 37)	6 9
AGE	ER_VIS	ER_VIS																		
>=19	>=6	<=10	8	11 43%	104	16 51%	(13 00)	60	20 20%	(7 50)	26	30 23%	(3 25)	26	37 14%	(3 25)	163	28 60%	(20 38)	6 3
AGE	ER_VIS	ER_VIS																		
>=19	>=11	60	5	7 14%	198	31 43%	(39 60)	156	52 53%	(31 20)	17	19 77%	(3 40)	16	22 86%	(3 20)	81	14 21%	(16 20)	4 8
	TOTALS		70	100 00%	630	100 00%	(9 00)	297	100 00%	(4 24)	86	100 00%	(1 23)	70	100 00%	(1 00)	570	100 00%	(8 14)	6 6

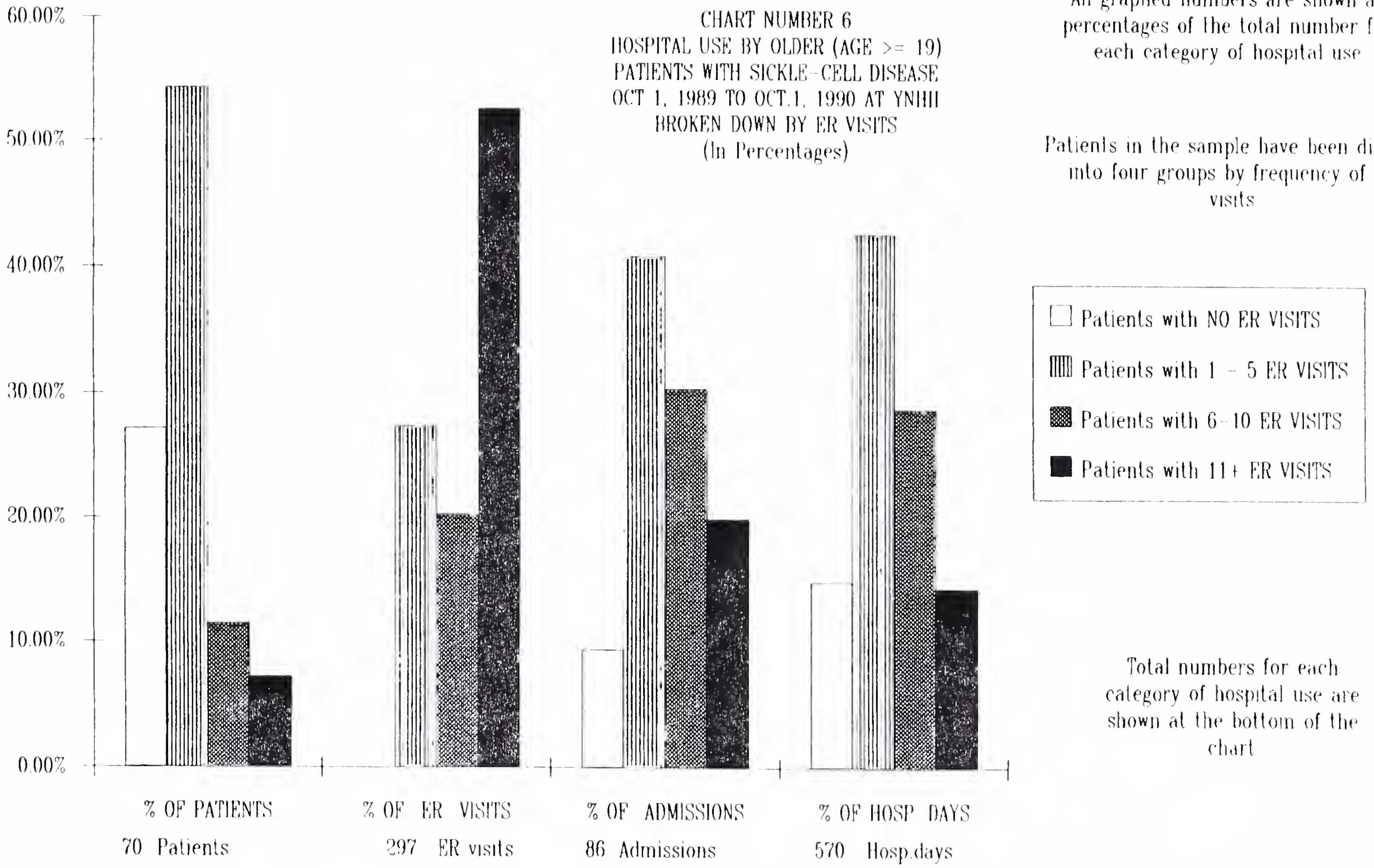


CHART NUMBER 6
HOSPITAL USE BY OLDER (AGE >= 19)
PATIENTS WITH SICKLE-CELL DISEASE
OCT 1, 1989 TO OCT.1, 1990 AT YNHH
BROKEN DOWN BY ER VISITS
(In Percentages)

All graphed numbers are shown as percentages of the total number for each category of hospital use

Patients in the sample have been divided into four groups by frequency of ER visits

Total numbers for each category of hospital use are shown at the bottom of the chart

Douglas Fleming, YMS

TABLE 7
(Corresponds to charts 7-10)

SICKLE CELL PROJECT YALE NEW HAVEN HOSPITAL

PATIENTS SEEN BETWEEN 10/1/89 AND 10/1/90

BREAKDOWN OF ER VISITS AND ADMITS FOR SICKLE CELL PATIENTS BY MONTH (AND BY TEMPERATURE)

TOTALS FOR ALL PATIENTS

	COLD MONTHS (TEMP < 40)								WARM MONTHS (TEMP > 60)			
	Oct '89	Nov '89	Dec '89	Jan '90	Feb '90	Mar '90	Apr '90	May '90	Jun '90	Jul '90	Aug '90	Sep '90
AVERAGE DAILY TEMPERATURES FOR EACH MONTH IN F	59	52	33	28	38	35	44	56	61	72	76	71

AGE	SEX	pt num		Oct '89	Nov '89	Dec '89	Jan '90	Feb '90	Mar '90	Apr '90	May '90	Jun '90	Jul '90	Aug '90	Sep '90
>=0	A	132	ER Visits	19	38	47	28	37	30	32	41	40	29	39	42
			Admits	8	20	19	17	11	13	11	7	15	11	15	17
			Temp (F	59	52	33	28	38	35	44	56	61	72	76	71

	4 COLD MONTHS (TEMP < 40)	4 WARM MONTHS (TEMP > 60)	TOTAL (12 MONTHS)
TOTAL ER VISITS	142	150	422
TOTAL ADMITS	60	58	164

TOTALS FOR ALL PATIENTS 19 AND OVER

AGE	SEX	pt num		Oct '89	Nov '89	Dec '89	Jan '90	Feb '90	Mar '90	Apr '90	May '90	Jun '90	Jul '90	Aug '90	Sep '90
>=19	A	70	ER Visits	13	25	33	18	28	23	24	31	30	20	23	29
			Admits	3	13	9	10	4	8	6	4	10	4	7	8
			Temp (F	59	52	33	28	38	35	44	56	61	72	76	71

	4 COLD MONTHS (TEMP< 40)	4 WARM MONTHS (TEMP >60)	TOTAL (12 MONTHS)
TOTAL ER VISITS	102	102	297
TOTAL ADMITS	31	29	86

TOTALS FOR ALL PATIENTS 18 AND YOUNGER

AGE	SEX	pt num		Oct '89	Nov '89	Dec '89	Jan '90	Feb '90	Mar '90	Apr '90	May '90	Jun '90	Jul '90	Aug '90	Sep '90
<=18	A	62	ER Visits	6	13	14	10	9	7	8	10	10	9	16	13
			Admits	5	7	10	7	7	5	5	3	5	7	8	9
			Temp (F	59	52	33	28	38	35	44	56	61	72	76	71

	4 COLD MONTHS (TEMP<40)	4 WARM MONTHS (TEMP >60)	TOTAL (12 MONTHS)
TOTAL ER VISITS	40	48	125
TOTAL ADMITS	29	29	78

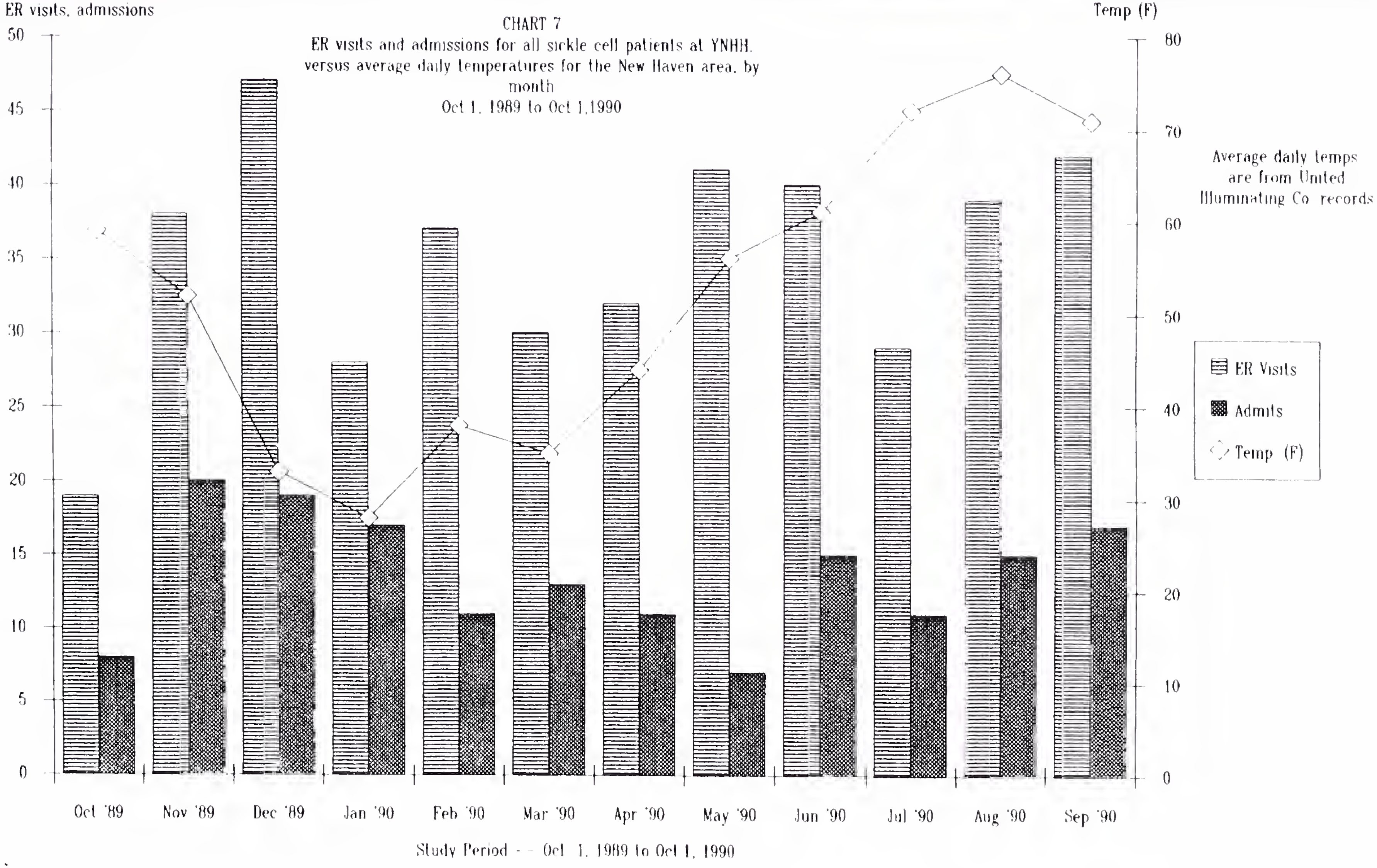
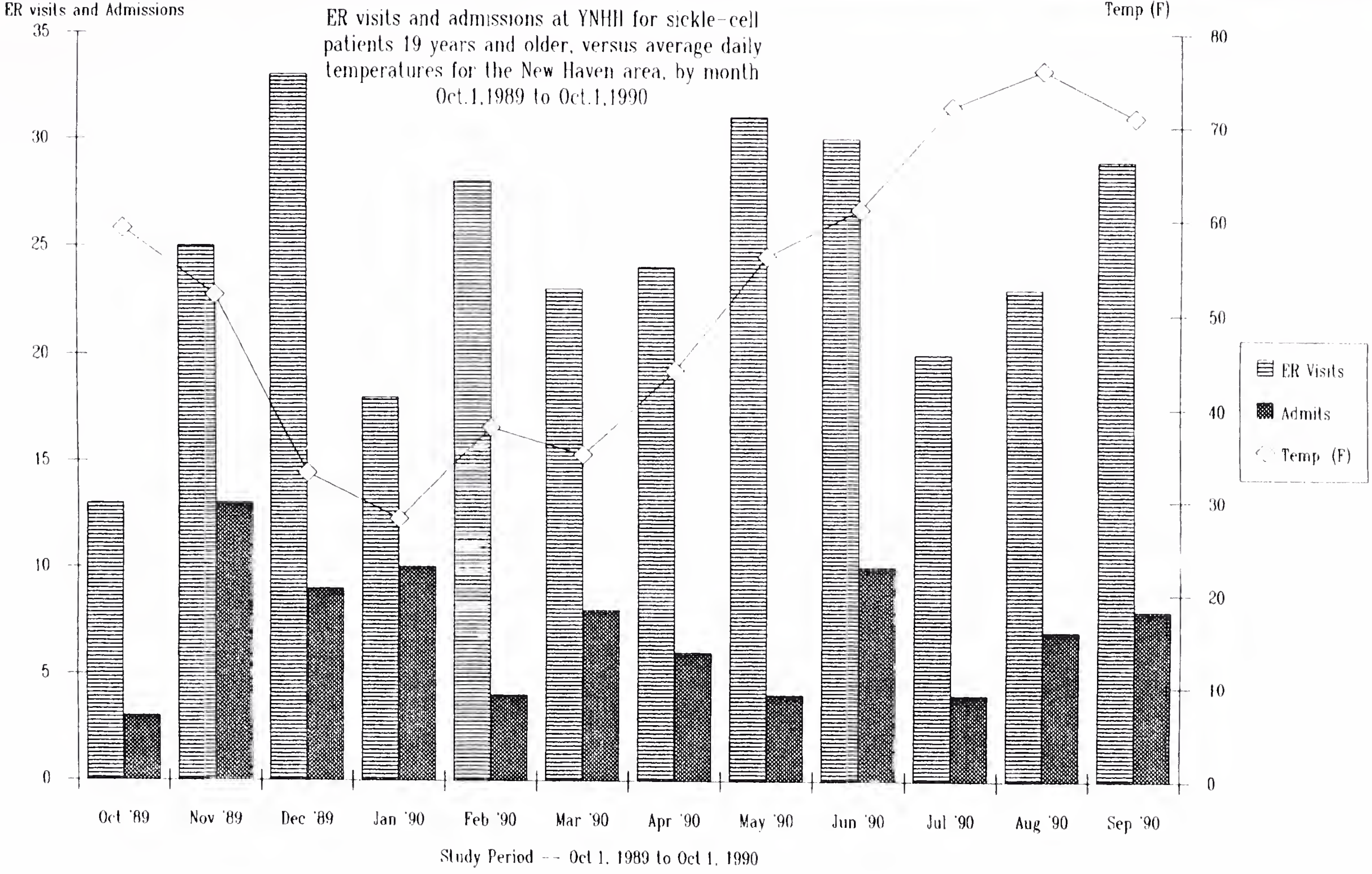


CHART 7
ER visits and admissions for all sickle cell patients at YNHH. versus average daily temperatures for the New Haven area. by month
Oct 1. 1989 to Oct 1,1990

Douglas T. Fleming. YMS

CHART 8

ER visits and admissions at YNHH for sickle-cell patients 19 years and older, versus average daily temperatures for the New Haven area, by month Oct.1,1989 to Oct.1,1990



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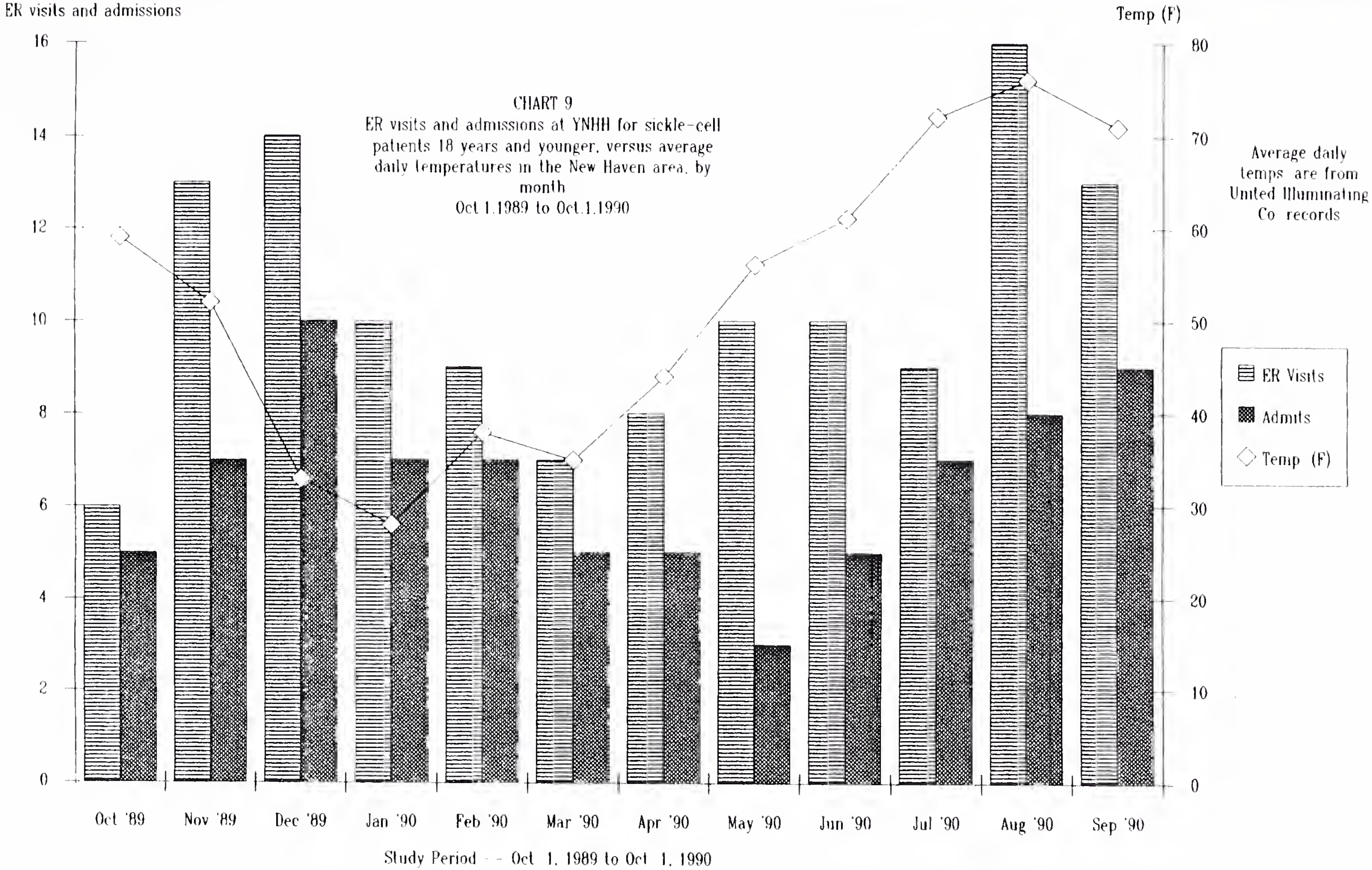


CHART 9
ER visits and admissions at YNHH for sickle-cell patients 18 years and younger, versus average daily temperatures in the New Haven area, by month
Oct 1.1989 to Oct.1.1990

Douglas T. Fleming, YMS

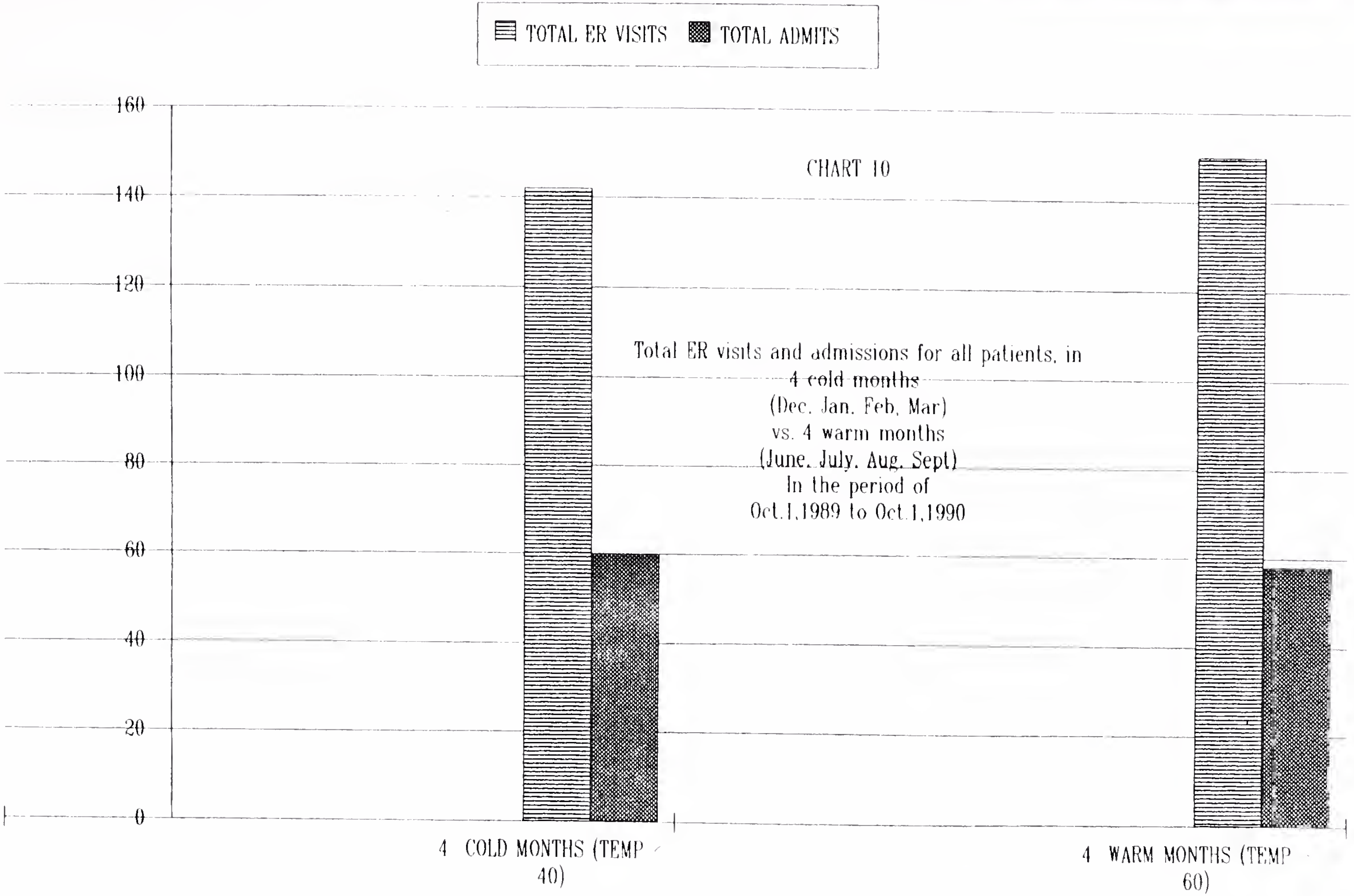
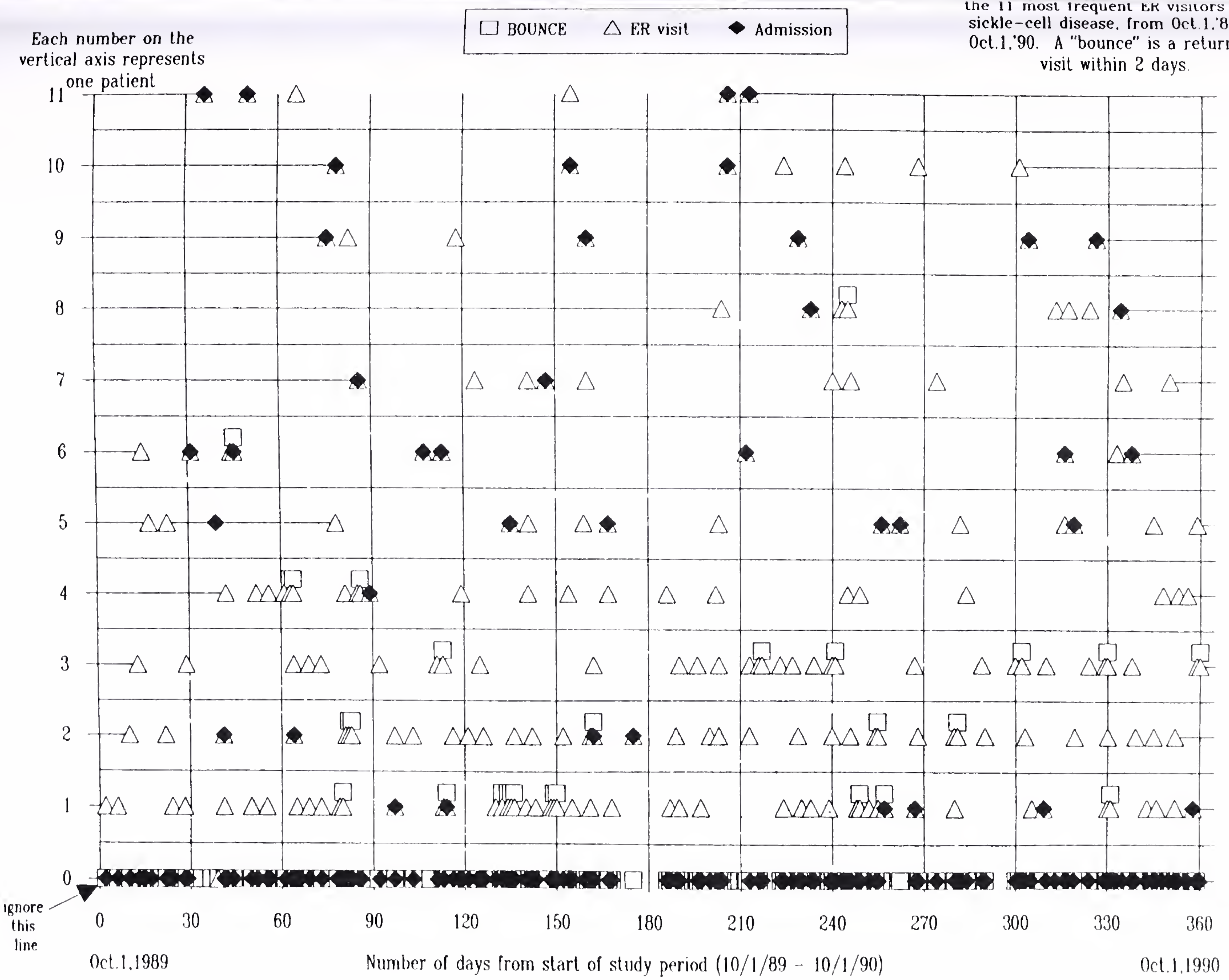


CHART 10

Total ER visits and admissions for all patients, in
4 cold months
(Dec. Jan. Feb. Mar)
vs. 4 warm months
(June. July. Aug. Sept)
In the period of
Oct.1,1989 to Oct.1,1990

Douglas T Fleming, YMS



the 11 most frequent ER visitors with sickle-cell disease, from Oct.1,'89 to Oct.1,'90. A "bounce" is a return ER visit within 2 days.

B. Phase 2 results

1.) Where does the patient receive his or her care?

All 8 of the interviewed patients stated that Yale-New Haven Hospital was the place they usually went when they wanted to see a doctor about their sickle-cell disease (Q2). When asked whom they would call first in the event of a sickle-cell crisis (Q3), all 8 patients replied that they would call the hematologist they knew from a Yale Hematology Clinic; 6 of the patients attended adult hematology clinic, and 2 attended pediatric hematology clinic.

Four of the adult patients had been seen at other health-care facilities in the past year (Q5): two patients had sought treatment in the Emergency Department of a nearby community hospital in New Haven, 3 times and 1 time respectively. In addition, two other patients had been out of state in the past year during a sickle-cell crisis, and had been hospitalized out of state, one time each. However, none of the patients had seen a private physician (that is, one with his or her own office outside a hospital) in the past year (Q6).

2.) Patient satisfaction with overall care

Patients were asked to evaluate the care they received in three areas of Yale-New Haven Hospital, the Hematology Clinics, the Emergency Department, and the inpatient floors of the hospital (ie while admitted). For each of these three areas, patients were asked to give a rating in three topics:

1) "In general, how well do the doctors ... explain to you what is going on with you medically?"

2) "In general, how well do doctors ... do as far as taking enough time to see you?"

3) "In general, how well do doctors ... do at treating the pain of your sickle-cell crises?"

Patients were asked to give a rating in each of these topics from among the following choices: 1) Poor, 2) Fair, 3) Good and 4) Excellent. In order to facilitate comparison of patients' responses in different hospital areas, grades of "excellent" were recorded as a score of "4", "good" as a score of "3" and so on. A response of "fair to good" was recorded as "2.5". In addition to the answers to individual questions, a combined score was calculated by adding together the ratings in the three topics, and then dividing by 3. Thus, the combined scores had the same possible range as the answers to individual questions, ie from 1 to 4.

Mean scores in the three topics for the 6 patients treated on the adult side of the Clinics, Emergency Department and inpatient floors were as follows, with standard deviations shown in parentheses ():

Hospital areas:

Topics:	Clinic	ED	Inpatient floors
1) Explaining:	3.2 (0.8)	2.2 (1.0)	3.5 (0.5)
2) Taking enough time:	3.0 (0.8)	2.2 (0.8)	3.3 (0.8)
3) Treating pain crisis:	3.3 (0.6)	2.2 (0.8)	3.7 (0.5)
4) Combined score	3.1 (0.5)	2.2 (0.6)	3.5 (0.5)

Because the sample size (6) was small, and there was no reason to expect that the variances of the scores in the three areas (Clinic, ED and admissions) would be different, the Student's T-test was used to compare average scores by the 6 patients treated as adults.

Patient's scores of quality of patient care in all areas were significantly lower for the Emergency Department than they were for the clinic or for the floors. As regards "treating pain crisis", for example, the ED received significantly lower ratings than the clinic ($P<.05$) and the inpatient floors ($P<.005$). Overall scores were also significantly lower for the ED than for the clinic ($P<.025$) and the inpatient floors ($P<.005$). Differences in scores between the clinic and the floors were not statistically significant.

It can be concluded that patients treated on the adult side of the hospital were significantly dissatisfied with the care they received in the

Emergency Department compared with the clinics and the floors. Overall score for the ED was 2.2, which was closest to "fair". Care in the clinics and inpatient areas received higher ratings of 3.1 and 3.5 respectively, which translate to between "good" and "excellent".

With only 2 patients in the sample treated on the pediatric side, it is difficult to come to any general conclusions regarding pediatric care. However, it is worth noting that both patients gave for the most part scores of 4 ("excellent") to the clinics, ED, and floors equally.

Patients' experiences in three areas of the hospital -- clinics, ED and inpatient -- will now be discussed separately.

3.) Experiences with Hematology Clinic

All six sample patients treated on the adult side of the hospital reported generally good long-term relationships with their clinic doctors at the Adult Hematology Clinic. Relationships with attending doctors were often of very long duration; in fact, one patient had been seeing her clinic attending for some decades. Four of the six patients had seen their clinic attending for 5 or more years. The shortest relationship was 2 years. Five out of the six patients gave their clinic attending a rating of "good" or "excellent" in "understanding their medical problems" ([128]).

In spite of these generally good ratings, 4 out of the 6 patients reported that they had had problems obtaining prescriptions at clinic in the past year. One of these patients reported that clinic doctors had been prescribing

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[128] Unfortunately, no question specifically addressed the differences in patients' perceptions of their attending physicians and of the hematology fellows, who also worked in clinic.

generic meperidine instead of the proprietary Demerol, and that the generic version did not relieve pain to the same degree. The patient ascribed this change to financial reasons: "My doctors at clinic say that Demerol is too expensive, and that my medical care [Title 19] won't cover it".

The other three patients who reported difficulty in obtaining pain medications felt that suspicions of drug-seeking or drug addiction were the source of the difficulty. In several instances, patients acknowledged having had actual problems around the issues of narcotics in the past, as in this quote:

> "If I'm going through a crisis, it can last 4 to 5 days. They used to give me 20 Demerol [tablets], and I'd have a few extra. So I'd sell them to another patient with sickle-cell disease. I was honest with my doctors at clinic -- I told [my doctor] myself, but they held it against me."

However, regarding the issue of addiction, the same patient noted that "when you're in pain, you just need something to help you -- that's not addiction. They [doctors] have never understood the pain, or experienced it."

More commonly, patients who experienced difficulty obtaining pain medications at clinic felt that they were encountering the suspicions generated by other patients, who had abused or sold their pain medications. For example, one patient remarked that

> "When I first came to clinic, they [the doctors] thought I was one of those people who sell their meds. Once they realized that I wasn't one of those people, everything was OK."

Similar echoes can be heard in this remark by another patient:

> "At first it was definitely slack -- it was a problem of people abusing pain killers, so the clinic doctors were real tight about giving pain killers. Now it's gotten better, at least for me. But other patients are still having problems."

Thus, these patients felt that establishing a trusting relationship with a physician in clinic was possible even if it was made difficult by the presence of other patients who abused pain medications.

One patient described an occasion when he had difficulty obtaining pain medications early in the course of a sickle crisis which later was complicated by a serious infection:

> "At first, [my doctor] didn't believe that the pain was so bad. He didn't want to give a lot -- this way he remained closer in contact. In the end, he was fair."

Thus, the physician's reluctance to give prescriptions for large amounts of pain medications seemed reasonable to the patient, as a way to stay "closer in contact".

It was possible to have the opposite problem as well, as one patient put it:

> "Clinic doctors don't hesitate to prescribe narcotics. There's no hesitation -- sometimes they even push the Demerol on me when I feel I don't need it."

Patients had a number of suggestions on how they felt clinic should be improved: for example, one patient felt doctors in clinic should not stick too inflexibly to "certain protocols they have for pain ... there are times when they should be more lenient, when a patient really needs it". Also, the

doctors should not be "so hurried". One patient remarked on the very long wait before a patient could be seen: "When you come to clinic, you have to wait and wait and wait for a doctor to see you. When you're in pain, the waiting is the hard part."

Overall, patients were generally positive about the care they received in adult clinic. As one patient remarked, "Overall, they're pretty good, the doctors I deal with. I like my attending doctor too."

The two patients treated in the pediatric hematology clinic gave glowing reports, such as this one:

> "They are excellent at clinic -- when I call, they get everything ready for me, my fluid bags and my medications. They even arrange everything for me when I go to the ER. They really know me and understand my problems."

Of course, no generalizations can be made about pediatric clinic due to the small sample size.

4.) Experiences with the Emergency Department

Interviews with six patients who were frequent visitors to the adult side of the Emergency Department during the study period revealed considerable conflict and anger around issues of pain relief and narcotic analgesics. In addition, patients remarked that they often felt neglected or forgotten in the hectic atmosphere of the Emergency Department. All six patients reported that there was no doctor at the Emergency Department who was responsible for their care on an extended basis, from visit to visit.

a.) Difficulty obtaining sufficient analgesia

When asked whether they had had any difficulty obtaining sufficient painkiller during visits to the ED in the last year, 5 out of the 6 patients replied that they had, and that the reason had been that "doctors did not believe that the patient needed more painkiller" (Q.20). The frequency of this problem was described as "very often" by 3 patients, "fairly often" by 1 patient, and "only seldom" by 1 patient. Interestingly, the 3 sickle-cell patients who answered that they had had difficulty obtaining painkiller "very often" were also the same sickle-cell patients who had visited the ED with the greatest frequency during the study period.

b.) Common themes in the interviews with adult patients

(1) Incomprehension of sickle pain

A number of common themes were brought up by patients in the interviews. Perhaps the most common was the feeling (cited by 4/6 patients) that there was little comprehension of the severity of sickle-cell pain among Emergency Department staff. In a particularly illustrative quote, one patient asked

> "How can the ER doctors know when someone isn't in pain? They don't understand how much pain sickle-cell patients have."

In another example, a patient remarked that

> "...sometimes the doctors tell you, 'the pain in your wrists isn't so bad -- go home and take some Tylenol.' But I say, 'if I could take some Tylenol at home, why would I come in here?'. It really makes me mad sometimes."

Thus, the patient was angry that appearing in the ED for pain relief was apparently not assumed by ED staff to be sufficient evidence that the pain could not be handled at home with non-narcotic medications. This feeling was echoed by another patient who had had difficulty obtaining sufficient pain relief: "I wouldn't go to the ED if I wasn't in pain."

(2) Accusations of drug-seeking and malingering

Patients frequently reported that they felt accused of drug-seeking, drug abuse, or malingering by ED staff (5/6 patients). As one patient put it, "they look at you like you're some kind of junkie". The accusation of drug abuse could take direct or indirect forms. As an example of a direct accusation, one patient noted that "one time a nurse made a smart remark -- she said I was on drugs, and that I had overdosed [before]". Another patient described a direct accusation of drug abuse as follows:

> "They treat you like dirt if they think you're injecting drugs. They accused me of shooting up and carrying drugs on me. I thought, now it's going to be hell getting treated. After that, they gave me only one shot [of Demerol], and I couldn't get a hold of my hematologist for more."

Thus, the patient felt that the accusation of drug abuse was directly connected to the withholding of narcotic analgesics. Note that the patient looked to his hematologist, that is, to his primary physician from hematology clinic, for help in obtaining treatment with narcotic analgesics.

Indirect accusations of drug-seeking were also described by several patients, and appeared to be more subtle. One patient described this type of interaction as follows:

> "People tell you that you're abusing drugs in a very indirect way -- they'll just hint around the issue for a while. Or they'll tell you about other people who abuse drugs, and make you feel like you're being lumped in with them."

Patients generally were quite taken aback by the accusation of drug-seeking, which they found to be unwarranted. For example, one patient remarked that "I've even been insinuated as being a drug user -- no one in the ED knows me or cares". This same patient reported that it had been necessary to get a "letter of reference" from a hematology fellow attesting to the patient's honesty and documenting the doses of Demerol previously needed to treat a crisis.

Several patients (2/6 patients), however, suggested that doctors were justified in some cases in being "careful" about analgesics, as in this remark: "Of course, they [the doctors] do try to cut down on painkillers, with all the abuse of painkillers going on". Another patient made a very similar remark: "There are some people who sell prescriptions, so doctors have to be very careful."

Two patients admitted to having had actual problems with drug use in the past. One patient had once attempted to self-medicate sickle pain by injecting a crushed Demerol tablet, and subsequently had a seizure on the way to the Emergency Department. Another patient had used "drugs" in an unspecified manner some 4 to 5 years previously, but stated that drug use had stopped. Both patients maintained that though they were not abusing narcotics presently, they felt that lingering suspicions raised by these previous incidents continued to obstruct their access to narcotic analgesics.

In a particularly interesting comment, one patient described the mode by which suspicions about drug-seeking were passed among physicians in the Emergency Department:

> "Some ER doctors treat you well. For others, some nurse will tell them something about me which changes their attitude completely -- a nurse might say, 'I saw him last week, and I don't think he's really sick'... Attitudes about me get passed from doctor to doctor, so that there are times when no one believes me when I need more painkiller. And then I might not be able to reach my hematologist, and can't get the relief I need, and it feels like I'm banging my head against the wall."

Thus the patient felt unable to escape previously aroused suspicions about drug-seeking, even when being treated for the first time by a new physician.

(3) Issues of power and control

Another important theme which emerged from several of the interviews (2/6) was the theme of power relations between physician and patient. One patient, for example, made the following remark:

> "There's one female doctor there [at the ED] who feels that she decides how much painkiller I get -- other people will ask me how much I need, but not her".

Thus, for the patient, the interaction between the patient and the physician in this case was centered around the control of medications. In another example, a patient who had had difficulty with substance abuse several years previously felt that pain medication was being withheld in a punitive fashion:

> "I need like 125 mg of Demerol and 50 of Vistaril [when I have a crisis]. When they won't give me as much as I need, they're punishing me -- I'm there for my condition... They look at me like they're thinking, 'you just want the drugs'".

This feeling of helplessness and punishment was echoed by another patient, who stated, "when they [the doctors] get you in a spot, they can really drive the pain in."

(4) Toradol

Two patients specifically mentioned the use of Toradol (generic name Keterolac), a non-steroidal anti-inflammatory medication. One patient reported that it had been tried twice, but had not been effective:

> "it just burned and made me sick in my stomach. I think they [the doctors] should have explained to us about it before experimenting on us."

In the case of another patient, the patient believed on one occasion that keterolac had been secretly substituted for meperidine, as the patient noticed no effect from the injection given. In addition, the patient was suspicious because the nurse had not gone to the narcotics room for medication prior to the injection. Thus, both patients who reported being treated with keterolac felt that this medication was ineffective, and that it was used in a deceptive fashion in their cases.

It should be noted that an extensive literature search revealed no study which documented the effectiveness of keterolac in sickle-cell disease.

c.) Interviews with 2 patients treated by the pediatric service of the Emergency Department

Only two of the patients who were interviewed had been treated on the pediatric side of the Emergency Department. Both of these patients were 23 years old, and would normally have been triaged to the adult medical service. However, they continued to be treated by the pediatric service, where they had been seen for many years by a special sickle-cell service overseen by pediatricians and coordinated by a nurse-practitioner. Both expressed reluctance to be transferred to the adult service because of the rushed atmosphere they perceived to exist there, as in this quote:

> "[My nurse] has told me that soon I'll have to go over to the adult ER -- but the patients on the adult side don't get the same treatment. The doctors over there don't know me, and they rush you through and don't give you the same attention as on the pediatric side."

Both patients seen by the pediatric service, therefore, were aware that their age made it more and more difficult to be retained by the pediatric service in the Emergency Department, but persisted in their attempts to avoid what they perceived to be a hectic atmosphere on the adult service.

d.) Overall impression of patients' experiences with the Emergency Department

The overall impression left by the 6 interviews with patients treated by the adult service of the Emergency Department was one of distrust and anger. These patients, who had all visited the Emergency Department frequently for sickle-cell crisis, felt that suspicions of drug-seeking directed against sickle-cell patients were prevalent (though not universal) among their physicians in the ED, and that this distrust, transmitted among physicians and nurses, made it difficult to receive sufficient narcotic analgesics. It is interesting to note that the two patients who were treated by the pediatric service were of an age normally treated by the adult service; both expressed reluctance to be transferred to the adult service because of the rushed atmosphere and lack of personal attention they perceived to exist on the adult service.

It must be understood that whatever basis ED staff had for their suspicions of drug-seeking, it could not be argued that frequent visitors to the ED did not suffer from very debilitating, serious illness. In fact, the eight patients interviewed were admitted to the hospital relatively frequently throughout the study year (an average of 3.6 times per patient). In addition, all eight patients had had at least two serious complications of sickle-cell disease in previous years. Among the eight patients, these complications included transient ischemic attacks or cerebrovascular accidents, pneumonias or pulmonary infarcts, cholelithiasis, hepatitis B, priapism, cardiac dilatation, ischemic heart disease, bony infarcts, pulmonary embolism, vertebral osteomyelitis, aplastic crisis, iron overload and transfusion resistance.

5.) Experiences on inpatient floors

The 6 patients who were hospitalized as inpatients on the adult medical service during the study period reported generally positive experiences. As noted above, patients treated on the adult service gave the inpatient floors a combined rating of 3.5, which translates to "good to excellent". Only 2 patients reported having had any difficulty obtaining sufficient pain relief, and both of these patients stated that this problem had occurred only "seldomly". One recurrent problem, reported by 2 patients, was that the waiting time in the Emergency Department before transfer to an inpatient floor could be extended, as long as 3 to 4 hours. Once on the inpatient floors, however, patients were generally satisfied with their care.

Two patients were hospitalized as inpatients on the pediatric service. Both were satisfied with the care they received, and neither reported any difficulty obtaining adequate pain relief for sickle crisis while admitted.

6.) Compliance with doctor's orders

Of the 8 patients interviewed, seven reported that they had been advised by their physicians to cut down on activities such as sports and heavy work, and all 8 reported that they had been advised to drink large quantities of water and to take other steps to avoid sickle crisis. As reported by patients, compliance was good, with all 8 patients stating that they followed their physician's advice either "always" (5 patients) or "usually" (3 patients).

Two patients noted that they occasionally played sports or participated in other strenuous activities against which they had advised by their physicians, but that playing sports helped them to stay fit and remain "like a normal person". Their decision to participate in strenuous activities was a personal one, and they did not express any conflict with their physicians over it. As one patient put it,

> "I felt that sports helped me, even if it brought on a crisis sometimes -- I try to stay active. Strenuous work causes crisis, so I follow doctor's orders not to do it."

What was notable, therefore, in patients' answers to questions about compliance was that there was little conflict with their physicians over steps to be taken to avoid sickle crisis. Steps such as drinking large quantities of water and avoiding strenuous effort, though burdensome at times, were generally seen by patients as reasonable.

7.) Disability

Of the 8 patients interviewed, all reported that sickle-cell disease had had a profound effect on their lives. The effects of the disease were greatest on patients' ability to earn a livelihood; all 8 patients reported that they would have liked to "work at a job or business" in the previous year, but that sickle-cell disease had prevented them from doing so at some time. Patients described the impact of the disease on their ability to work at a job or business as "very large" (7 patients) or "large" (1 patient). Of the 8 patients, 5 were on disability, one was "temporarily out of work", one was "not usually employed", and one worked part time.

Effects of the disease on patients' ability to continue their education were also great. Seven of the eight patients stated that they would have liked to "go to school" in the previous year, and all of these 7 patients reported that sickle-cell disease had prevented them from doing so at some time. When these 7 patients were asked to describe the impact of the disease on their ability to continue their education, 4 patients responded with "very large", one with "large", one with "moderate", and one with "small". Of the total 8 patients, one had completed a college education, 3 had started college but not graduated, and 4 had graduated from high school.

In contrast, the impact of sickle cell disease on patients' ability to keep house and do household chores did not appear as great. Five of the 8 patients reported that their illness had prevented them from "keeping house" at some time in the previous year, but the impact of the illness in this area was judged to be "small" or "moderate" by 4 out of these 5 patients.

Patients mentioned periodic absences due to sickle crisis as a reason for their difficulty with employment and with education, as in the following remark:

> "I can't get the type of job I want -- I need more training, but I miss classes when I get sick, and I get behind."

Patients expressed frustration that in spite of periods of relatively good health, recurrent pain crisis made regular employment or education very difficult. This frustration was typified in the following remark:

> "I always wanted to be like my sister and brother, but I couldn't -- I'm much more dependent on my parents than they are. I really want to start working, so that I can be more on my own. It's hard when you're always sick."

In general, therefore, patients considered themselves to be significantly disabled, especially in employment and education. Painful sickle crisis was not just a periodically troublesome phenomenon in their lives, but a source of more general difficulty. Though psychological issues of self-esteem and dependence were not explored more deeply in the interviews, remarks such as the one just cited make it clear that difficulties with employment and education have wide-ranging psychological ramifications.

It should be pointed out that the patients interviewed were all frequent visitors to the Emergency Department, and that their experieneces with disability may not reflect the disability of sickle-cell patients in general.

8.) Access to medical care

Patients reported generally good access to medical care. Of the 8 patients interviewed, 7 had Medicare or Title 19 assistance, and 1 had insurance with a private insurer. Yale-New Haven Hospital was the medical-care facility preferred by all 8 patients; none had been forced to seek care at Yale because financial constraints had prevented them from being seen elsewhere.

Two of the patients had had difficulty in the past year paying for medical care. In one case, a dispute with a private insurance company led to difficulty paying, and in another case, a patient whose Medicare paid 80% of hospital bills had difficulty paying the remaining 20%. Only one patient reported that a financial difficulty had actually impaired care. In this case, the patient reported that because Title 19 would not pay for proprietary Demerol, it was necessary to purchase the less expensive generic version, Meperidine. Though the patient found generic meperidine to be ineffective, the effect of this substitution was still judged to be "mild".

These findings about access to health care among frequent visitors to the Emergency Department will be important in considering the patient-physician relationship. It is important to emphasize in any case that it was not generally financial pressure which forced patients to seek care at YNHH rather than at nearby community hospitals. Conversely, from the hospital's standpoint, it is important to note that even though only one of the eight patients interviewed was working regularly (part-time), all had some form of insurance, primarily governmental (Title 19, Medicare or both for 7 of the 8 patients). Thus, care to these frequent ED visitors was not being supplied `gratis' by the hospital.

9.) Patients' overall perspective on the medical care system

In the final question of the interview, patients were asked to answer the following question, designed to assess overall perspective on the medical care system:

> Which of the following statements comes closest to expressing your overall view of the American health care system? (1g)
>
> 1) (Poor) The American health care system has so much wrong with it that we need to completely rebuild it.
>
> 2) (Fair) There are some good things in our health care system, but fundamental changes are needed to make it work better.
>
> 3) (Good) On the whole, the health care system works pretty well and only minor changes are necessary to make it work.

Perhaps somewhat surprisingly, patients appeared to have a generally favorable perspective on the medical care system. Of the eight patients interviewed, five patients answered "3", and three patients answered "2". Thus the majority of these patients, with their life-long exposure to the health care system, remained convinced that "the health care system works pretty well and only minor changes are necessary to make it work".

V. Discussion

The unifying hypothesis of the present study, first presented in the "Introduction", may now be examined in light of the study's results. The hypothesis posits that a small subset of sickle-cell patients with especially severe symptoms becomes known to the Emergency Department staff as visiting the ED frequently and requiring large doses of narcotic analgesics for painful crisis. Irrespective of the actual prevalence of drug dependence among patients, suspicions of drug dependence may arise, leading to a tendency among ED physicians to limit the prescription of narcotic analgesics for sickle-cell patients in general. Underprescription of pain medication may lead to requests for additional pain medication by patients, or to repeat ED visits, and thus in turn to escalating suspicions of drug-seeking among ED staff. In this way, suspicion of drug-seeking among ED staff and frustration at lack of pain relief among patients may begin to fuel one another in a continuing circle.

The present study aimed to investigate this hypothetical circle at several points, and to gather the information necessary to organize management interventions to interrupt it. Therefore, the first task at hand was to survey the ED and other hospital services for use by patients with sickle-cell disease. A computer survey identified all visits to the ED by sickle-cell patients over a one-year period (1 October 1989 to 1 October 1990).

The computer survey identified a total of 132 patients with sickle-cell disease, with a mean age of 21 years. Of the 422 visits to the Emergency Department accounted for by sickle-cell patients during the study year, 131 (31.0%) resulted in admission. These 131 ED admissions represented 79.9%

of the 164 total admissions for the group. Admissions overall lasted an average of 5.9 days each.

Results of the computer survey largely confirmed the hypothesis that small numbers of very frequent visitors to the Emergency Department accounted for a disproportionate number of visits. Among patients with 6 or more ED visits during the study year, 6 younger patients (10% of all younger patients) accounted for 50% of all ED visits by younger patients, whereas 13 adult patients (19% of all adult patients) accounted for 73% of ED visits by adults. These results are consonant with previous studies of ER utilization by "frequent visitors" with sickle-cell disease ([129]).

Results of the interviews with 6 adult patients who visited the adult medical service of the Emergency Department frequently (6 or more times during the study period) confirmed the hypothesis that considerable conflict between patients with sickle-cell disease and physicians may develop over issues of drug-seeking and drug dependence, especially in the Emergency Department. Five of the six patients reported that they had experienced difficulty obtaining sufficient pain relief during crisis at some time in the last year, because "doctors did not believe that the patient needed more painkiller". Patients reported that they felt that ED staff did not comprehend the pain they experienced during crisis. In addition, patients frequently felt accused by ED staff, directly or indirectly, of drug-seeking or drug abuse. These problems were not reported by patients to the same degree with the hematology clinics or with the inpatient floors.

Two of the 8 patients interviewed, though both 23 years old, continued to be seen by the pediatric service. These two patients gave equally high ratings to the pediatric ED, hematology clinic and inpatient floors. Though their number is too small to permit generalizations, it is interesting to note

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[129] See, for example, Vichinsky (1982), p.329, Powers (1986). Also of interest are Pierce (1990) and Selbst (1990).

that both patients expressed reluctance to be transferred to the adult medical service of the Emergency Department, where they felt that a rushed atmosphere prevailed, judging from conversations with adult sickle-cell patients. It is important to note that pediatric sickle-cell patients at YNHH are cared for by a special sickle-cell service overseen by pediatricians and coordinated by a nurse-practitioner.

Time considerations did not permit interviews with sickle-cell patients who visited the Emergency Department a moderate amount, for example, between 1 and 5 times per year. These patients represent the great majority of patients in the community with sickle-cell disease, and it would be very useful to compare their experiences in the Emergency Department with those of more "frequent visitors". As it is, it is not known whether conflicts which develop between ED staff and "frequent visitors" also affect the care received by the great majority of sickle-cell patients. Follow-up studies will investigate this hypothesis. However, it is reasonable to suppose that conflicts with these "frequent visitors" may color perceptions among ED staff of sickle-cell patients in general.

Patients reported that the physicians who saw them in hematology clinic, namely, hematology fellows and attendings, had a pivotal role in managing their care. Patients described long-term relationships with their hematology attendings (from 2 years to decades), and reported generally positive experiences in hematology clinic. Physicians from hematology clinic frequently visited patients when they sought care in the Emergency Department, and often interceded for patients in assisting them to obtain adequate pain relief in the Emergency Department.

The Emergency Department physicians with whom patients described the most conflict were almost universally members of the hospital housestaff. In this context, it is important to note that the frequently exhausted housestaff bear the brunt of caring for patients on the "front lines", and are

most directly subject to the frustrations of dealing with patients one-on-one. At the same time, they necessarily have limited experience in dealing with knotty issues such as possible drug-seeking and dissembling by patients. In addition, it is reasonable to suggest that housestaff, as young physicians, may be uncomfortable in discovering the acute limitations of their profession in curing the sick ([130]). In all these respects, the experience of housestaff is in contrast with that of attending physicians (and, for that matter, hematology fellows), who are somewhat protected from the stresses of daily life in the ED, and who may be more comfortable dealing with the uncertainty and limitations of clinical medicine.

It should be noted that Yale-New Haven Hospital is the only medical facility in its area with a significant experience and expertise in treating hematological diseases such as sickle-cell disease. The closest academic hematology training programs outside of YNHH are in New York City, Boston, or Farmington, Connecticut, all a considerable distance from New Haven. Though patients in crisis in the New Haven area could also seek care at nearby community hospitals, few of the patients interviewed actually did. In fact, all 8 of the interviewed patients denied that they would have preferred to be seen in some other hospital. This situation might have been different in a larger city, in which patients who were dissatisfied with their care in one Emergency Department could seek care at a variety of other Emergency Departments at hospitals with expertise in hematology.

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[130] See esp. Mechanic (1968) on issues of housestaff vs. attending physicians.

VI. Suggestions for improving care of sickle-cell disease

A. General considerations

From the standpoint of the Emergency Department, sickle-cell disease is both similar to a number of other illnesses, and entirely unique. For example, sickle-cell disease might be compared to other chronic, debilitating illnesses with periodic exacerbations, such as asthma, diabetes, hemophilia, migraine headaches or lower back pain. In each of these illnesses, as in sickle-cell disease, patients may visit the Emergency Department with high frequency over many years, and may present difficult problems of diagnosis and treatment. However, sickle-cell disease distinguishes itself from these illnesses by its intractability, its severe pain symptoms requiring treatment with narcotic analgesics, and in the constant possibility of life-threatening complications with each pain crisis. Thus, Emergency Department treatment of sickle-cell disease faces relatively unique difficulties.

A number of possible steps might contribute to alleviating the present situation of tension and conflict in the Emergency Department. Any program to address the issue of acute care of sickle-cell patients must consider the following points:

1) Though the literature suggests that the great majority of sickle-cell patients do not become dependent on pain medications, drug-seeking and drug dependence are possible among patients with sickle-cell disease, given the chronic nature of sickle pain and the need for narcotic analgesics.

2) Patients with sickle-cell disease are often significantly disabled, especially in employment and education, and have an increased prevalence of psychosocial difficulties.

3) The possibility of serious medical complications exists during every painful crisis. Early detection of complications such as osteomyelitis, for example, may greatly reduce final morbidity and shorten the hospital course.

B. Suggestion: better coordination of care for sickle-cell patients

For all these reasons, coordination of the care of each patient with sickle-cell disease -- in the psychological, psychosocial, and medical realms -- is pivotal. At present, hematology fellows and attendings coordinate the care of individual sickle-cell patients in the medical realm, but do not have the resources coordinate care for the patient in the Emergency Department, nor to coordinate psychological and psychosocial aspects of care. Sickle-cell patients who seek care at the Emergency Department on a given day may be able to speak with their primary hematology attending, but may also have to be seen by whichever of the many hematology attendings and fellows is on call that day. At present, there is no one person acquainted with sickle-cell patients over the long term, who is able to act as a sort of "central clearinghouse" in the management of their care.

One model for coordinating the care of patients with sickle-cell disease is close at hand, and already operating on the pediatric service. Among pediatric sickle-cell patients, care is organized by a special sickle-cell service overseen by pediatricians and a coordinated by a nurse-practitioner. Within this system, the special needs of sickle-cell patients can be addressed in an

organized fashion, in the outpatient hematology clinics as well as in the Emergency Department.

C. Suggestion: Management Protocols for pain management

At the level of the Emergency Department, the use of individualized management protocols would assist in standardizing the care of sickle-cell patients who frequently utilize emergency services, and in reducing confusion and conflicts among different services over treatment of individual patients. Such individualized management protocols, drawn up in consultation with each patient and his or her hematologist, and updated as needed, would provide a short summary of the patient's history, specify the type and starting doses of analgesics to be given for sickle-cell crisis, and list the names and telephone numbers of physicians responsible for each patient's care. Nurses and house-staff would be instructed to consult the treatment protocol prior to the initiation of any therapy for sickle-cell patients presenting for pain, and to contact the patient's primary physician during each ED visit.

In fact, the use of such management protocols has been sporadically attempted in the Emergency Department of Yale-New Haven Hospital. Approximately two years ago (ie approx. 1989), according to ED staff, management protocols emphasizing the use of oral pain medications and limiting the use of IM injections were instituted for several of the most frequent ED visitors with sickle-cell disease. However, as described by ED staff, such protocols were unsuccessful in reducing patients' requests for medication and repeat visits for pain relief. This lack of success was attributed to the difficulty in enforcing such protocols, due to the fragmented nature of care for sickle-cell patients and the day-to-day changes in the hematologist on call. Thus, from the standpoint of ED staff, the key factor in

the failure of such management protocols was the lack of a collective will to enforce them, among ED staff and members of the Hematology Section.

It is also possible, in the hectic atmosphere of the Emergency Department, that confusion over when and where to find such management protocols may have contributed to their failure. One approach to this problem would be to program the ED computer system to print a copy of the management protocol at the time that each sickle-cell patient registers.

A pivotal point in designing a program to coordinate the care of sickle-cell patients is that the object is improved care, and not merely decreased use of IM narcotic pain medications or decreased numbers of repeat visits. It is imperative that patients in pain crisis receive adequate analgesia. The design of a successful program will involve not only ED physicians, hematologists, and nurses, but most importantly, the patients themselves. As such, the design of each management protocol should take place with active participation by the patient, in order to make the protocol a part of therapeutic alliance, rather than an obstacle to receiving pain relief.

As was discussed in the literature review, success with management protocols in reducing conflicts over drug-seeking and in reducing frequency of ED visits has been reported in several studies at other institutions. In Powers' study (1986), 5 men with sickle-cell disease were identified as chronic ED users; with these 5 patients, a management protocol was associated with a reduced total number of ED visits (from 163 to 56 in 6-month periods before and after institution of the program), reduced mean length of ED stay, and reduced use of parenteral meperidine, as well as increased satisfaction with care among both patients and ED staff. In Vichinsky et al's study (1982), a multidisciplinary, comprehensive pain program, which included a management protocol for treatment of pain, was associated with a 58% reduction in Emergency Department visit and a 44% decrease in pain-related hospitalizations in 10 patients with sickle-cell disease.

Management protocols drawn up even for a small subset of sickle-cell patients at Yale-New Haven Hospital could have a large impact on their care. Data from the present study indicate, for example, that only 5 patients at Yale-New Haven Hospital accounted for nearly 53% of all ED visits by sickle-cell patients over the study year. Thus, management protocols drawn up for these 5 patients, if consistently and humanely applied, could reduce confusion and conflict in over half of the 420-some visits to the ED by sickle-cell patients over the course of a typical year. One may be confident that reduction in conflict over the treatment of these five "frequent visitors" will result in improved care for other sickle-cell patients as well.

VII. Bibliography

1.) **Abbuhi** S, Jacobson S, Gibson G: "Serum concentration of meperidine in patients with sickle cell crisis", in Annals of Emergency Medicine 15:433-438 (1986). Cited in Shapiro, 1989, p. 1035.

2.) **Angell** M, "The Quality of Mercy", NEJM 306, 98-99 (1982)

3.) **Barrett**, DH, et al, "Assessment of Psychosocial Functioning of Patients with Sickle-Cell Disease", Southern Medical Journal, vol.81, no. 6, (June 1988)

4.) **Baum**, KF, Dunn, DT, and Maude, GH, et al: "The Painful Crisis of Homozygous Sickle-Cell Disease", Arch. Intern. Med. 147:1231-1234 (1987)

5.) **Bobo**, L, et al, "Health Perceptions and Medical Care Opinions of Inner-City Adults with Sickle-Cell Disease or Asthma Compared with Those of Their Siblings", Southern Medical Journal, vol. 82 no. 1, pp. 9-12. (Jan, 1989)

6.) **Bonica**, The Management of Pain, 2nd edition. Lea and Febiger, Pennsylvania. (1990)

7.) **Brodner**, RA and Taub, A., "Chronic Pain Exacerbated by Long-Term Narcotic Use in Patients with Non-malignant Disease", Mount Sinai J. Med. 45:233, (1978) (cited Bonica, 1990, p. 598)

8.) **Brownell**, A, Lowson, S, and Brozovic, M, "Serum ferritin concentration in sickle cell crisis", J. Clin. Path, 39, 253-255

9.) **Brozovic**, M, Davies, S, and Yardumian, A, et al, "Pain Relief in Sickle Cell Crisis", (letter). Lancet 2:624-625 (1986) (shows no addiction) (cited Shapiro, 1989)

10.) **Bunn** HF, "Disorders of Hemoglobin", in Harrison's Principles of Internal Medicine, 11th ed., McGraw Hill, pp. 1519-1523 (1987)

11.) Samuel **Charache**, "The Treatment of Sickle Cell Anemia", Arch Intern Med 133:698-705 (1974). Cited Barrett, 1988, p. 746.

12.) Samuel **Charache**, "Treatment of Sickle-Cell Anemia", Ann. Rev. Med 32:195-206 (1981)

13.) Stephen **Cole** and Robert Lejeune, "Illness and the Legitimation of Failure", American Sociological Review, Vol. 37, pp. 347-356 (1972). Reprinted in Howard D. Schwartz, Dominant Issues in Medical Sociology, 2nd ed, New York: Random House, pp. 31-40.

14.) **Cole** T. Sprinkle R, Smith S, et al: "Intravenous Narcotic Therapy for Children with Severe Sickle Cell Pain Crisis", Am J Dis Child 140:1255-1259 (1986) (withholding analgesics ---> illicit use) (cited Shapiro, 1989) See also reply by Mallouh below.

15.) C. Lockard **Conley**, "Sickle-Cell Anemia -- The first Molecular Disease", in Wintrobe, MM, ed., Blood, Pure and Eloquent. New York: McGraw Hill, 1980.

16.) **Damlouji** NF, Georgopoulos A, Kevess-Cohen R, et al: "Social Disability and Psychiatric Mobidity in Sickle Cell Anemia and Diabetes Patients", Psychosomatics 23:925-931 (1982) (cited in Shapiro, 1982 -- ordered through interlibrary loan)

17.) **Diggs** LW, Ching RE: "Pathology of Sickle-Cell Anemia", Southern Med. J. 27:839-845, (1934) (cited in Conley, p. 334)

18.) **Fordyce** WE: Behavioral Methods for Chronic Pain and Illness, St. Louis, C.V Mosby, 1976. Cited in Bonica, 1990, p. 602.

19.) **Fordyce** WE, et al: "Acute Back Pain: A Control Group Comparison of Behavioral vs. Traditional Management Modes", J. Behav. Med. 9:127 (1986). Cited in Bonica, 1990, p.1462-64.

20.) **Freeman** HE, 1986 National Survey of Access to Health, Principal investigator Howard E. Freeman, #8981. Or, see a report of the study's results in Freeman, H.E., Blendon, R.J. et al, "Americans Report on Their Access to Health Care" in Health Affairs, Spring 1987, Vol. 6, no. 1, 6-18)

21.) **Goldberg** MA et al, "Treatment of sickle cell anemia with hydroxyurea and erythropoeitin", NEJM 323(6):366-72 (Aug.9,1990)

22.) **Hahn** EV, Gillespie EB: "Sickle-cell Anemia. Report of a case greatly improved by splenectomy. Experimental study of sickle cell formation." Arch Intern Med 39:233-254 (1927). Cited in Conley, p.332

23.) **Harris** D, "An Elaboration of the Relationship between General Hospital Bed Supply and General Hospital Utilization", J of Health and Social Behavior 16: 163-172 (cited in Waxler, 1981, p. 90)

24.) **Heller** P, Koshy M, Schade S: "Analgesics in Sickle Cell Anemia", JAMA 260:703-704 (1988) (cited Shapiro, 1989)

25.) **Herrick**, JB: "Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia", Arch. Intern. Med. 6:517-521 (1910). Cited in Conley, p. 320.

26.) **Hurtig** AL, White LS: "Psychosocial adjustment in children and adolescents with sickle-cell disease", J. Pediatric Psychology 11:411-427 (1986). Cited in Shapiro, p. 1033.

27.) **Ingram** VM, Scientific American 198:68-74. Cited in Conley, p. 346

28.) **Kumar** S, Powars D, Allen J, et al: "Anxiety, Self-Concept, and Personal and Social Adjustments in Children with Sickle-Cell Anemia", J Pediatr 8:859-863 (1976)

29.) **Lawrence** C, Fabry ME: "Objective Indices of Sickle-Cell Painful Crisis: Decrease in RDW and percent dense cells and increase in ESR and Fibrinogen." Prog. Clin. Biol. Res. 240:329-336 (1987). Cited in Shapiro, 1989, p. 1031.

30.) Sandra R. **Leavell**, Charles V. Ford, "Psychopathology in patients with Sickle-Cell Disease", Psychosomatics, vol.24, no.1, p. 23-37 (Jan. 1983)

31.) Deborah B. **Leiderman**, Jean-Anne Grisso, "The Gomer Phenomenon", Journal of Health and Social Behavior, vol. 26, pp 222-231 (1985). Reprinted in Howard D. Schwartz, Dominant Issues in Medical Sociology, 2nd ed, New York: Random House, pp. 287-296

32.) **Lemanek** LK, Moure SL, Gresham FM, et al: "Psychological Adjustment of Children with Sickle Cell Anemia", J. Pediatr. Psych 11:397-409 (1986). Cited in Shapiro, 1989, 1033, #57.

33.) **Lessin** LS, Jensen WN: "Sickle Cell Anemia 1910-1973", Arch Intern Med 133:529-532 (1974)

34.) **Ludwig** E, and Adams S, "Patient Cooperation in a Rehabilitation Center: Assumption of the Client Role", J. Health and Social Behavior 9:328-336. (cited in Waxler, 1981, p.91)

35.) Nicole **Lurie**, et al, "Termination from Medi-Cal -- Does it affect health?", in NEJM 311:480-484 (Aug. 1984). See also a follow-up article by Lurie et al, "The termination of Medi-Cal Benefits -- A follow-up study one year later" in NEJM 314:1266-1268 (May. 1986)

36.) **Mallouh** A: "Intravenous Narcotics in Sickle Cell Crisis", (letter), Am J Dis Child 141:1039-1040 (1987) (argues against use of narcotics because of potential for abuse -- cf Cole, 1986) (cited Shapiro, 1989)

37.) **Mankad** VN, Williams P, Harpen M, et al: "Magnetic Resonance Imaging, percentage of dense cells and serum prostanoids as tools for objective assessment of pain crisis: a preliminary report", Proc. Clin. Biol. Res. 240:337-350 (1987). Cited in Shapiro, 1989, p. 1031.

38.) David **Mechanic**, Medical Sociology, 2nd ed, New York: Free Press, esp. pp. 407-432

39.) Jane **Mercer**, Labelling the Mentally Retarded, berkeley: University of California Press, 1973, (cited in Waxler, 1981)

40.) Solomon **Papper**, "The Undesirable Patient", J. of Chronic Diseases vol 22:777-779 (1970). Reprinted in Howard D. Schwartz, Dominant Issues in Medical Sociology, 2nd ed, New York: Random House, pp.85-101.

41.) Talcott **Parsons**, The Social Contract

42.) Linus **Pauling**, "Abnormality of hemoglobin molecules in hereditary hemolytic anemias" in Harvey Lect., 1953-1954, series 49. New York, Academic Press, 1955, pp. 216-241. Cited Conley, 1980, p. 339

43.) **Pearson** H et al: "Developmental Pattern of Splenic Dysfunction in Sickle Cell Disorders", Pediatrics 76, p. 392-397 (1985)

44.) **Pearson** H: "Sickle cell syndromes and other hemoglobinopathies" in Miller DR, Baehner RL, McMillon CW et al (eds): Blood Diseases of Infancy and Childhood, ed. 5. St. Louis, Mosby Co. (1984) pp. 402-438.

45.) **Perlin** E, Castro O, Finke H, et al: "The value of diflunisal (Dolobid) in the treatment of painful sickle cell crisis", Blood 68:S64a, 1986.

46.) John M. **Pierce**, Arthu L. Kellerman, Catherine Oster: "'Bounces': An Analysis of Short-Term Return Visits to a Publin Hospital Emergency Department", Annals of Emerg. Med. 19:752-757 (July, 1990)

47.) Orah S. **Platt**, "Is there Treatment for Sickle-Cell Anemia?", NEJM 319, 1479 (Dec. 1, 1988)

48.) Russel K. **Portenoy** and Kathleen M. Foley, "Chronic Use of Opioid Analgesics in Non-malignant Pain: Report of 38 Cases", Pain 25, 171-186 (1986) (report on long-term trial of opioids in 30-some patients with a wide variety of illnesses, mostly low back pain -- no sicklers were included. Cited Shapiro 1989, footnote # 71)

49.) **Powers** RD: "Management Protocol for Sickle-Cell Disease Patients with Acute Pain: Impact on Emergency Department and Narcotic Use", Am J Emerg Med 4:267-268 (1986) (cited in Shapiro, 1982)

50.) **President's Commission** for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research, "Screening and Counseling for Genetic Conditions", p. 20, February 1983.

51.) **Purdie** FRJ, Honigman B, Rosen P: "The chronic emergency department patient" Annals of Emergency Medicine 10:298-301 (1981)

52.) **Ready**, LB, Sarkis, E, and Turner, JA, "Self-reported Versus Actual Use of Medications in Chronic Pain Patients". Pain 12:285 (1982) (cited Bonica p. 598)

53.) Derek **Robinson**, "Sickle Cell -- Point, Counterpoint", NEJM 289:323-4 (1973) (discusses the public outrage - esp. in the black community - over mandatory screening for SCD in newborns and school children. See letter by Rutkow below)

54.) Ira M. **Rutkow** and Jeffrey M. Lipton, "Mandatory Screening for Sickle-Cell Anemia" (letter), NEJM, 289,865 (1973) (a letter giving data to confirm Robinson's remarks on the dangers of mandatory screening of newborns and school children for SCD)

55.) Alan N. **Schechter** and H. Franklin Bunn, "What determines severity in Sickle Cell Disease?" (editorial) in NEJM, 306:295-297 (1982)

56.) **Schechter** NL, Berrien FB, Kanz SM: "The Use of Patient-controlled Analgesia in Adolescents with Sickle Cell Pain Crisis: A Preliminary Report", J Pain Sympt Manag 3:109-113 (1988) (cited in Shapiro, 1989 -- ordered through interlibrary loan)

57.) **Schuckman** A, Fontanarosa PB, "'Frequent Fliers' -- A protocol for the management of frequent visitors to a community hospital emergency department" (abstract) Annals of Emergency Medicine 18:435 (1989)

58.) Steven **Selbst**, Mark Clark: "Analgesic Use in the Emergency Department", Annals of Emergency Medicine 19:1010-1013 (Sept. 1990)

59.) **Serjeant** GR: Sickle Cell Disease, Oxford, Oxford University Press, 1985.

60.) Barbara S. **Shapiro**, "The Management of Pain in Sickle-Cell Disease", Pediatric Clinics of North America, vol. 36 no.4 (Aug 1989)

61.) G. **Stamatoyannopoulos**, "Problems of Screening and Counseling in the Hemoglobinopathies", in A. Motulsky and W. Lenz, eds, Birth Defects" Procedings of the Fourth International Conference (Vienna, 1973), Medica, Amsterdam, 1974

62.) **Szeto** H, Inturrisi CE, Houde R, et al: "Accumulation of normeperidene, an active metabolite of meperidine, in patients with renal failure of cancer", Annals of Internal Medicine 86, 736-741 (Oct 1977).

63.) **Tang**, R, Shimomura S, Rotblatt M, "Meperidine-induced seizures in sickle cell patients", Hospital Formulary 76:764-772, (Oct. 1980) Cited in Shapiro, 1989, p. 1035, and Cole, 1986, p. 1259.

64.) Elliott P. **Vichinsky**, Robert Johnson, and Bertram H. Lubin, "Multidisciplinary Approach to Pain Management in Sickle-Cell Disease", American Journal of Pediatric Hematology-Oncology, vol. 4 no. 3, (Fall, 1982)

65.) **Waddell** G, et al: "Non-organic Physical Signs in Low Back Pain", Spine 5:117 (1980). Cited in Bonica, 1990, p.1457-58.

66.) Nancy E. **Waxler**, "The Social Labelling Perspective on Illness", Originally in Arthur Kleinman and Leon Eisenberg, eds, The Relevance of Social Science for Medicine, Dordrecht, Holland: D. Riedel Publishing Company. Reprinted in Howard D. Schwartz, Dominant Issues in Medical Sociology, 2nd ed, New York: Random House, pp.85-101.

VIII. Appendix: Survey Questions

Sickle-Cell Project -- Douglas Fleming, Yale Medical Student

Revision date: 2 February 1991

Check when entered into the computer: ○

Face Sheet for Interviews with Patients

Patient name code: Gender: Age:

Sent letter? Yes / No

Phone number code:

On hand: 1) extra pencils 2) watch
Check: 3) "Do not disturb" sign 4) pagination 5) sent letter
6) visit-list or chart for confirmation of sickle disease

Date and time of first attempt at phone contact:

○ Successful, did interview

○ Reached patient, but pt asked to be called later
(will call back on date: time:

○ Reached pt, but pt did not want to be interviewed
○ No answer ○ Busy

Date and time of second attempt at phone contact:

○ Successful, did interview

○ Reached patient, but pt asked to be called later
(will call back on date: time:

○ Reached pt, but pt did not want to be interviewed
○ No answer ○ Busy

Date and time of third attempt at phone contact:

○ Successful, did interview

○ Reached patient, but pt asked to be called later
(will call back on date: time:

○ Reached pt, but pt did not want to be interviewed
○ No answer ○ Busy

Date and time of fourth attempt at phone contact:

○ Successful, did interview
○ Reached patient, but pt asked to be called later
(will call back on date: time:
○ Reached pt, but pt did not want to be interviewed
○ No answer ○ Busy

Sickle-Cell Project -- Douglas Fleming, Yale Medical Student

Faculty advisor: Joel Rappeport, MD

Survey of Patients with Sickle Cell Disease:

What questions will be asked.

Hello, my name is ____________, and I'm calling from Yale Medical School. We are doing a survey of people with Sickle-cell Disease who go to Yale-New Haven Hospital, regarding their experiences with the health-care system. I'd like to ask you some questions about your health experiences and medical care. You are free to answer or not to answer -- your participation is entirely voluntary, and will not affect the care you receive. Would you be willing to spend a few minutes answering some questions?

(If patient agrees) First, I will ask some general questions about your experiences with health care, and then I will ask you some questions about your last visit to Yale-New Haven Hospital. All of your answers will be strictly confidential. This means that none of your doctors or other hospital staff will be able to find out how you answered any question. Also, you can decide at any time not to answer any question, or to stop answering questions altogether. The interview will last about 20 minutes.

(Numbers in parentheses indicate the question number of the corresponding question in the 1986 National Survey of Access to Health, Principal investigator Howard E. Freeman, #8981. Or, see a report of the study's results in Free, H.E., Blendon, R.J. et al, "Americans Report on Their Access to Health Care" in Health Affairs, Spring 1987, Vol. 6, no. 1, 6-18)

*** Q.1 Would you say your health, in general, now is.... (1)

1) Poor 2) Fair 3) Good 4) Excellent

<<<<< Where does the pt receive care? >>>>

*** Q.2 Is Yale-New Haven Hospital the place where you usually go when you want to see a doctor (about your sickle-cell disease in general)?

1) Yes 2) No

*** Q.3 Which doctor would you usually call or go to first when you have a sickle-cell crisis?

1) A doctor from Yale adult hematology clinic
2) A doctor from Yale Pediatric heme clinic (Dr. Pearson and Suzanne et al)
3) a doctor from a clinic at Saint Raphael's Hosp.
4) a doctor from a clinic at Bridgeport Hosp.
5) a doctor from a clinic at Hill Health Center
6) A private doctor
7) An HMO
8) Other: ________________________
9) YNHH **Emergency Room**
10) Some other **Emergency Room** (which one?____________)

*** Q.4 Is your sickle-cell disease followed over time by a hematologist or hematologists, that is, by a doctor who specializes in blood diseases?

a) 1) Yes 2) No

b) (if YES) Where does this hematologist work?

1) Yale adult hematology clinic
2) Yale pediatric hematology clinic (Dr. Pearson, Suzanne et al)
4) Saint Raphael's
5) Bridgeport
6) Hill Health Center
3) A hematologist in private practice (in his or her own office)
4) A hematologist working with an HMO
5) Other hematologist

*** Q.5 Is there any other hospital or clinic where you have gone for medical care of your sickle-cell disease in the last year, that is, since (present month) of last year?

a) 1) Yes 2) No

b,c) (If YES) Which one is that? (circle all that apply)

1) Saint Raphael's
2) Bridgeport
3) Hill Health Center
4) Other hosp. ____________
5) Other hosp. ____________

d) How many times have you been seen at this hosp/clinic (s) in the past year? ________

*** Q.6 Have you seen a private doctor (in his own office, not in a hospital) about your sickle-cell disease in the past year?

a) 1) Yes 2) No

b) (If YES) Where does he or she practice?

c) (If YES) How many times have you seen this private doctor in the past year? _________

<<<< Hematology Clinic >>>>

*** Q.7 a) Have you been to the hematology clinic (pedi or adult) at YNHH in the past year, that is, since January of last year?
Y/N

b) Were you seen in the pediatric or the adult hematology clinic?
Pedi / Adult

c) When was the last time you went to Hematology Clinic at Yale, approximately? ________

(if pt has not been at YNHH hematology clinic (adult or pedi) in the past year, skip to the next section)

*** Q.8 When you visit the hematology clinic at Yale, is there one particular clinic doctor who manages your care as your primary doctor? (23b)

a) 1) Yes 2) No 3) Not applicable

b) (If Yes) How long has this doctor been seeing you in clinic?

____________________________ years/months/weeks

c) (If YES) How well does this doctor understand your medical problems?
1) Poor 2) Fair 3) Good 4) Excellent

*** Q.9 In general, how well do the doctors in clinic explain to you what is going on with you medically?
1) Poor 2) Fair 3) Good 4) Excellent

*** Q.10 In general, how well do the doctors in clinic do as far as taking enough time to see you?
1) Poor 2) Fair 3) Good 4) Excellent

*** Q.11 In general, how well do doctors in clinic do at treating the pain of your sickle-cell crises?
1) Poor 2) Fair 3) Good 4) Excellent

*** Q.12 In the past year, when you have visited the Hematology Clinic at Yale, have you had any difficulty obtaining a prescription for all the painkiller you needed?

a) 1) Yes 2) No 3) N/A

(If YES) What have been the sources of the difficulty? (check all that have been sources of difficulty in the past year)

b) Financial reasons -- pt could not afford to be seen (Y/N)

c) Doctors did not believe that pt needed more painkiller (Y/N)

d) Other reason for not receiving painkiller (Y/N)

e) (If YES to a)) In the past year, how often have you experienced this difficulty obtaining painkillers in clinic?

1) only seldomly 2) Fairly often 3) Very often

*** Q.13 Explain (comments on clinic experiences):

<<<<< Emergency Room >>>>>>

*** Q.14 a) Have you been to the Emergency Room at YNHH (pedi or adult) at YNHH in the past year, that is, since January of last year?
Y/N

b) (If YES) Were you seen on the pediatric or the adult side of the Emergency Room?
Pedi / Adult

c) When was the last time you went to the ER at Yale, approximately? ________

(if pt has not been at YNHH ER (adult or pedi) in the past year, skip to the next section)

*** Q.15 When you visit the Emergency Room at Yale, is there one particular emergency room doctor who manages your care as your primary doctor? (that is, one who works primarily in the ER, not someone, for example, from hematology clinic who comes down to the ER to see you) (23b)

a) 1) Yes 2) No 3) Not applicable

b) (If Yes) How long has this doctor been seeing you in the Emergency Room?

______________________ years/months/weeks

c) (If YES) How well does this emergency room doctor understand your medical problems?

1) Poor 2) Fair 3) Good 4) Excellent

*** Q.16 (If patient has a relationship with a Yale clinic doctor -- see answer to Q.4)
When you visit the ER at Yale, how often does your doctor from the hematology clinic come to see you in the ER?

0) N/A

1) Never 2) Sometimes 3) Usually 4) Always

*** Q.17 In general, how well do the doctors in the Emergency Room explain what is going on with you medically?

1) Poor 2) Fair 3) Good 4) Excellent

*** Q.18 In general, how well do the doctors in the Emergency Room do as far as taking enough time to see you?

1) Poor 2) Fair 3) Good 4) Excellent

*** Q.19 In general, how well do doctors in the emergency room do at treating the pain of your sickle-cell crises?

1) Poor 2) Fair 3) Good 4) Excellent

*** Q.20 In the past year, when you have visited the Emergency Room at Yale, have you had any difficulty obtaining a prescription for all the painkiller you needed?

a) 1) Yes 2) No 3) N/A

(If YES) What have been the sources of the difficulty? (check all that have been sources of difficulty in the past year)

b) Financial reasons -- pt could not afford to be seen (Y/N)

c) Doctors did not believe that pt needed more painkiller (Y/N)

d) Other reason for not receiving painkiller (Y/N)

e) In the past year, how often have you experienced this difficulty obtaining painkillers in the Emergency Room?

1) only seldomly 2) Fairly often 3) Very often

*** Q.21 Explain (comments on ER experiences):

<<<< Inpatient care >>>>

*** Q.22 a) Have you been admitted to the hospital at YNHH for sickle-cell disease in the past year, that is, since January of last year?
Y/N

b) Did you stay on the pediatric wards, or on the adult wards?
Pedi / Adult

c) When was the last time you were admitted at Yale-New Haven Hosp., approximately? ________________

(if pt has not been admitted at YNHH in the past year, skip to the next section)

*** Q.23 In general, when you are admitted at YNHH, how well do the doctors in the hospital explain to you what is going on with you medically?

1) Poor   2) Fair   3) Good   4) Excellent

*** Q.24 In general, when you are admitted, how well do the doctors in the hospital do as far as taking enough time to see you?

1) Poor   2) Fair   3) Good   4) Excellent

*** Q.25 In general, when you are admitted, do doctors and nurses in the hospital do well at treating the pain of your sickle-cell crises?

1) Poor   2) Fair   3) Good   4) Excellent

*** Q.26 In the past year, when you have admitted at Yale-New Haven Hospital, have you had any difficulty obtaining a prescription for all the painkiller you needed?

a) 1) Yes 2) No 3) N/A

(If YES) What have been the sources of the difficulty? (check all that have been sources of difficulty in the past year)

b) Financial reasons -- pt could not afford to be seen (Y/N)

c) Doctors did not believe that pt needed more painkiller (Y/N)

d) Other reason for not receiving painkiller (Y/N)

e) In the past year, how often have you experienced this difficulty obtaining painkillers while admitted overnight in the hosptal?

1) only seldomly 2) Fairly often 3) Very often

*** Q.27 Explain (comments on experiences while admitted):

<<<<<< Compliance >>>>>

*** Q.28 Have your doctors advised you to cut down on activities such as sports or heavy work because of your sickle-cell disease? (68f)

a) 1) Yes 2) No

b) (If YES) Are you following the doctor's instructions?

1) hardly ever or not at all
2) sometimes
3) usually
4) always

*** Q.29 Have your doctors advised you to drink a lot of water or take other steps to avoid sickle crises? (adapted from 69a)

a) 1) Yes 2) No

b) (If YES) Are you following the doctor's instructions?

1) hardly ever or not at all
2) sometimes
3) usually
4) always

*** Q.30 In general, do you follow the recommendations given to you by your doctors?

1) hardly ever or not at all
2) sometimes
3) usually
4) always

*** Q.31 (open question) Why or why not? (and in which ways)

<<<<< Disability >>>>

*** Q.32 In the last year, that is, since (this month) last year, has sickle-cell disease prevent you from...

a) Working at a job or business? (70b)
1) Yes 2) No 3) N/A
b) (If YES) How great an impact has sickle-cell disease had on your ability to work at a job or business?
1) small 2) moderate 3) large 4) very large

c) keeping house? (70d)
1) Yes 2) No 3) N/A
d) (If YES) How great an impact has sickle-cell disease had on your ability to keep house?
1) small 2) moderate 3) large 4) very large

e) Going to school? (70f)
1) Yes 2) No 3) N/A
f) (If YES) How great an impact has sickle-cell disease had on your ability to go to school?
1) small 2) moderate 3) large 4) very large

g) Carrying out your normal activities in any way? (70j)
1) Yes 2) No
h) (If YES) How great an impact has sickle-cell disease had on your ability to carry out your normal activities?
1) small 2) moderate 3) large 4) very large

*** Q.33 Are you currently.... (83a)
1) employed full time
2) employed part time
3) temporarily out of work
4) retired
5) not usually employed
6) on disability
7) keeping house/homemaker
8) other (specify:_________________

<<<<<<<< Pain medication >>>>>>>

*** Q.34 a) During the last year, how many months out of the year have you needed to take at least some pain medication (for sickle-crisis or for between-crisis pain)?

______ (answer from 1 to 12)

b) (Pain meds, cont'd) In the past year, how often have you needed pain medication at home, when you are not in the hospital?

1) Never or hardly ever
2) Occasionally (once a month or several months)
3) Fairly frequently (several times per week or several weeks)
4) All or most of the time (most days)

c) (If yes to b) When you have pain, which pain medication do you usually actually take for the pain you have at home?
(drug, dose and route) ____________________

d) (If yes to b) Which pain medication works best for this pain you have at home? (ie, which pain medication would you prefer to get?)
(drug, dose and route) ____________________

e) When you have a bad sickle-cell crisis, which pain medication do you prefer to get (and how much):

(drug, dose and route) ____________________

f) When you have sickle-cell pain crisis, how effective is IV narcotic medication in stopping the pain you feel?
0) Not effective at all
1) a little effective
3) moderately effective
4) very effective
(record N/A if pt has not tried this method)

g) When you have sickle-cell pain crisis, how effective is oral narcotic medication in stopping the pain you feel?
0) Not effective at all
1) a little effective
3) moderately effective
4) very effective
(record N/A if pt has not tried this method)

h) When you have sickle-cell pain crisis, how effective is non-narcotic medication in stopping the pain you feel?
0) Not effective at all
1) a little effective
3) moderately effective
4) very effective
(record N/A if pt has not tried this method)

i) comments on pain medication:

<<<<<<< Access >>>>>

*** Q.35 In general, would you prefer to go somewhere else other than Yale-New Haven Hospital ER or clinic to receive your care? (32a)

a) 1) Definitely yes 2) Perhaps 3) No

b) (for YES and PERHAPS only) Where would you have preferred to go? (32B)

____

c) (for YES and PERHAPS only) Why didn't you go there? (32e)

____

code answer: (all that apply)
1) Financial -- couldn't afford someplace else
2) Transportation
3) Health insurance or HMO covers visits only to YNHH
4) Regular doctor/HMO etc was not seeing patients
5) Referred by some other provider
6) Other reason

*** Q.36 How much difficulty have you had in the past year paying for your medical care at YNHH?

a) 1) No difficulty 2) Some difficulty 3) A lot of difficulty

b) (If 2 or 3) Why was that?

code answer:
1) Financial -- Had no insurance, couldn't afford payments for medical care
2) Financial -- Had health insurance or HMO, but this did not cover all costs
3) Other (will add codes as appropriate)

*** Q.37 In the past year, was there any time when the costs of medical care prevented you from getting all the care you needed (or felt you needed) at YNHH?

a) 1) Yes 2) No

b) (If YES) How much of an effect do you think this had the care you received?

1) mild effect 2) Strong effect 3) Severe effect

*** Q.38 Explain (open question):

<<<<<< BACKGROUND >>>>>>

*** Q.39 What is the highest year in your education which you have completed? (eg, in elementary school, high school, or college) (81)

0) None
1) Elementary -- 1 year
2) 2 years
3) 3 years
4) 4 years
5) 5 years
6) 6 years
7) 7 years
8) 8 years
9) High school 9 years
10) 10 years
11) 11 years
12) H.S. grad 12 years
13) College 13 years
14) 14 years
15) 15 years
16) College grad. 16 years
17) Some graduate school
18) Graduate or professional

*** Q.40 What is your marital status?

1) Single -- never married
2) married
3) divorced or separated
4) widowed

<<<< Overall Perspective >>>>

*** Q.41 Which of the following statements comes closest to expressing your overall view of the American health care system? (1g)

1) (Poor) The American health care system has so much wrong with it that we need to completely rebuild it.

2) (Fair) There are some good things in our health care system, but fundamental changes are needed to make it work better.

3) (Good) On the whole, the health care system works pretty well and only minor changes are necessary to make it work.

Time at this point:

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